UNIVERSITY OF BIRMINGHAM

**School of Education**

sense
for deafblind people

# The experiences of people with rare syndromes and sensory impairments in hospitals and clinics

## Executive Summary

Dr Liz Ellis, (University of Birmingham) Lucy Keenan (Sense), and Dr Liz Hodges (University of Birmingham)

21st October 2015

Funded by Sense

# Acknowledgements

We are grateful to many people who made this study possible; putting them in any kind of order is impossible. They were all essential.

Firstly however, we are very grateful indeed to all the participants who gave their time, both patients and their families, and were so ready to talk to us and allow us to learn something of their lives.

We are also grateful to the hospitals and the hospital staff who helped and supported us, most especially;

Professor Timothy Barrett, Birmingham Children's Hospital, who was our key medical contact and helped us navigate the ethics process

Our other principal investigators who gave us their time and support and allowed us to visit their clinics:
Dr Tarek Hiwot, Queen Elizabeth Hospital Birmingham
Dr Jane Valente, Great Ormond Street Hospital London
Mr Martin Snead, Addenbrooke's Hospital Cambridge
Professor Andrew Webster, Moorfields Hospital London

They all have more than enough to do without helping us. We are very grateful for their time.

Other clinical staff also supported us and allowed us to visit their clinics, we are grateful to all and especially to Mr Robert Henderson. We would also like to thank all the staff in hospital research departments who supported us in navigating sometimes complex systems.

We would like to thank all members of the advisory committee and especially Dr Isabel Clare for her support throughout. We thank Yemi Tadesse from Sense for her input in the early months. We are also grateful for the contributions of Dr Anna McGee from Sense for her support, advice and encouragements and Dr Graeme Douglas, University of Birmingham, who provided advice and inspiration in his support for the project.

# Executive Summary

This study was born from informal discussions with people with rare syndromes (particularly those with sensory impairments) who were describing difficulties they were having as **patients**. In many cases they were already carrying a large burden of problems; they had multiple, complex individual symptoms, and they were often aware their conditions could or would deteriorate; they were dealing with a world unfriendly to people with sensory impairments.

The research study was therefore set up to investigate the following issues;

- How do people with sensory impairments (and their families/carers) experience their attendance at clinics in relation to their rare syndromes?
- What are the factors in relation to clinic type, environment, and attitude that affect this experience?
- What makes the delivery of services efficient and effective for people with sensory impairments and their families, across and between clinics and what factors might inhibit this?

## What is good practice?

These Good Practice Guidelines are presented first, as they summarise what was learnt from the study. However, they were compiled from the information gathered from interviews, environmental data and accompanied visits. They are thus drawn from patient experience as expressed to us through the study, and emerged from the data, rather than being a pre-existing measure for the hospitals we visited and talked about.

These guidelines fall under three broad headings; Good practice in the environment, Good practice by staff, and Good practice in clinical situations.

They are divided into two sections, the first section includes items which might require authorisation, expenditure, systems reorganisation or other

large scale thought and work. The second section however requires much less in terms of infrastructure, although it might involve training, and is more concerned with simple, and inexpensive changes which can be undertaken by people who are prepared to make an effort.

## Good practice in the environment

*Policy strategies*

- Buildings are easy to navigate, using colour coding of areas, clear and consistent signage, multiple formats of information
- There is good even lighting throughout corridors and in cafés and toilets
- There is at least clarity about car parking – such as patients only having to pay for what they have consumed (such as payment on exit)
- Department names are kept the same throughout the site (so not both “eye clinic” and “ophthalmology”)
- Toilets for people who need to lay down to be changed are made available
- All consulting rooms to have four walls and a solid door

*Operational strategies*

- Reception staff should offer directions and support for wayfinding on arrival
- Lights are kept on and additional lighting is provided in dark areas
- More play/activity equipment is provided in waiting areas, particularly thinking about the needs of older children, and also children who require specific sensory toys. TVs with subtitles and signing, and wifi are made available.
- Deliberate and clear paths are kept across waiting spaces – perhaps marked with tape/paint as clear paths
- Toilet leaflets are provided - simple directions for how toilets work, which are offered to patients

## Good practice in staffing

*Policy strategies*

- All staff, including reception and nursing staff, are trained in issues around sensory impairment, communication techniques and guiding
- There is a review and assessment policy for each hospital which includes a walk through and overview of procedures and clinics – such as 15 Steps, with a focus on sensory impairment issues
- Liaison personnel are provided; non-medical staff who can point patients in the right direction for both medical information, procedural information, and further support
- Training for staff in understanding the issues of dilation (vision reduction) for all patients, but particularly those who are deaf/ have a hearing impairment

*Operational strategies*

- Use of a sticker or colour-coded protocol or similar which outlines patients' communication and information needs and which all staff ***read***. This is now a part of the Accessible Information Standard and will need to be implemented by July 2016
- All staff, reception and clinical, regularly check communication needs - 'Is that communication OK for you? Can I ask to you to repeat that so I know you understand?'
- Time is always given for genuine listening to patient – for those with rare syndromes only they can know the individuality of their conditions
- All staff always introduce themselves, clearly, including their job title, and this information is available in written (print, email, braille) format both before, and after appointments
- Staff give patients time, understanding that patients may have travelled a long way and have waited a long time
- Staff look at patients when talking, do not cover their mouths and pay full attention to patients during discussion
- Staff talk to patients, not interpreters or companions

- Staff ask if patients would like them to read information leaflets/forms/documents to them, and assist in filling out forms
- A culture of asking three key questions;
  - How can I help?
  - Am I getting this right?
  - What else can I do?

**Good practice in clinic practice; appointments and procedures**

*Policy strategies*

- Multiple formats of leaflets/information are always available (large print, audio, braille, electronic) and given to patients rather than leaving them to be noticed by patients
- Co-ordinated appointments are offered to minimise pressure on patients and families – either for families (syndromes often run in families) or multiple conditions (so that audiology and cardiology are carried out on the same day, or are on the same day for siblings)
- Procedures requiring anaesthesia are grouped to minimise the number of anaesthetics given
- Formal arrangements are made for patients to be able to meet by having ‘syndrome’ days for patient appointments
- Informal arrangements are encouraged for patients to meet each other – perhaps co-ordinated on social media, but a room is provided
- Loop systems are always available, working (checked) and used
- A quiet, private room is available for discussion with the receptionist, clinician or other

*Operational strategies*

- Appointment and post appointment information is available in accessible formats, for individual needs; always checking, is this format still Ok for you?

- Appointment and post appointment information is always available in digital formats if this is preferred
- Patients are given clear advance information about what the appointment is for; and what might happen as a result of it (patients were not always clear and some are juggling so many appointments, they have to prioritise)
- A plan of what exactly will happen at appointment or clinic is provided in advance, with who they will see, to minimise confusion
- Text messages are used to aid communication – e.g. to confirm appointments or tell people when to return from the café or outside if clinics are running late
- Information is promptly given in appropriate formats about waiting times; reasons for waiting; text messages are sent about delays
- There is a clear system for ensuring that people know when they are called to an appointment once in clinic, including approaching people directly if they need this
- Staff willingly talk to patients if they have difficulty with automated systems (e.g. booking in machines)
- Specialist clinics (e.g. ophthalmology, audiology) understand the needs of their own patients- e.g. vision impairment or hearing impairment
- There are smaller areas (with screens or similar) for children who find noise and bustle difficult
- Staff are ready to guide people to rooms when they are called and will ensure that someone will take them back
- Staff are always willing to provide, afterwards, written feedback on what was said in an appointment, in appropriate formats, especially digital information
- Named contact for further information following the clinic visit

## Patients, clinics and syndromes

We focused on six particular syndromes which frequently cause dual sensory impairments and which are all rare (fewer than one in 2,000 people have them). Amongst the considerations which led us to these groups were; that they all cause sensory impairments and frequently dual sensory impairments; that all had particular clinics or practitioners who focused on them; that several of them were ciliopathies; that there was a patient identity associated with conditions, related to, for example, the existence of a support group.

The syndromes we thus included were; Alström syndrome; Bardet-Biedl syndrome; CHARGE syndrome; Stickler Syndrome; Usher syndrome and Wolfram syndrome. We also included the experiences of 5 people with syndromes that were different to these six main syndromes.

**Wolfram syndrome**'s key factors are diabetes insipidus, diabetes mellitus, optic atrophy and deafness. It is a degenerative disorder. It affects 1 in 770,000 people in the UK.

**Alström syndrome** affects the whole body, including with rod-cone retinal dystrophy, sensorineural hearing loss, obesity, insulin resistance, and type 2 diabetes mellitus, as well as a range of other issues. It is a life-limiting disorder, which might affect 100 people in the UK .

**Stickler syndrome** is a group of conditions which affect connective tissue (collagen). Stickler is characterised by a distinctive facial appearance, cleft palate, eye problems, hearing loss, and joint problems. It affects about 1 in 8,000 people in the UK

**CHARGE syndrome**'s principal factors are with the eye, choanal atresia (when the nasal passages are blocked by bone or tissue), cranial nerve anomalies, and ear anomalies. They may have learning delay. It affects about 1 in 12,000 people in the UK.

**Usher syndrome** (type 1, 2 and 3) is principally characterised by sensorineural deafness and progressive vision loss due to Retinitis Pigmentosa (RP). Initially peripheral vision loss occurs which is described as 'tunnel vision'. The exact number of people affected is unknown but it could be as high as 1 in 7,000 people.

**Bardet-Biedl syndrome**'s main features are retinal degeneration, extra fingers and/or toes and obesity. Diabetes mellitus is also common. It affects about 1 in 160,000 people in the UK.

This study looked predominantly at three kinds of clinics;

**Specialised holistic services**; clinics provided yearly with a range of specialisms involved for a particular syndrome (Alström, Wolfram and Bardet-Biedl each have these type of clinics).

**Condition specific clinics**; a clinic in which a consultant and the team will focus on a particular condition (Stickler, Usher and CHARGE syndrome have these kind of clinics)

**General clinics**; clinics which are neither of the above but are still often highly specialist.

## Methodology

In the course of this project, over 20 months, we gathered information from 52 participants (either people with rare syndromes and sensory impairments or their families), from all over England, representing people from 14 months to 83 years old. We have collected many hours of interview data, including some in BSL, and attended 5 hospitals, incorporating 8 clinics, to see how things work in practice, where the clinics are situated, and sometimes alongside the people attending.

The study incorporated a number of information gathering tools

*Interview with patients*; initially patients with one of the six syndromes who attended specialist clinics were interviewed; later this expanded to other people with dual sensory impairments and with some other conditions who attended a range of clinics. The interviews investigated travel and mobility to and within the hospital, the information provided before and after the clinics, and communication from receptionists, clinical staff and others.

*Environmental audits*; these were carried out in five hospitals to provide additional data for comparison. They examined patient areas such as corridors, waiting areas, toilets and cafés, and clinic rooms.

*Accompanied visits*; we also accompanied five patients on their visits through hospitals, to gain some insight into how long procedures took, what communication methods staff used, how aware staff were of the needs of people with sensory impairments, the time given to communicate, including reading papers, and more.

The study gained ethical approval from the NHS (NREC) and from each of the participating hospitals. Patients and their families were asked for their consent, using appropriate formats considering age, communication method, and ability. 52 people were interviewed and told us of their experiences of rare syndromes, clinics, and sensory impairments; of these, 42 were people who have a rare syndrome, and the remaining 10 were from parents/guardians of people with a rare syndromes speaking about the experiences of clinics for the people they care for and in their own role as a parent or carer.

## The patient experience;

From the exploration of the experiences of participants we analysed the data gain from the three research tools to draw out points in relation to key moments in the patient's journey through the clinic; appointments, in the clinic, the consultation, and after the clinic.

### Appointment

Patients talked about issues around getting an appointment, and the different routes to this depending on the kind of clinic they were attending. They considered specialist clinics and clinicians to be very valuable, although distances to clinics and the length of time between referrals could be a problem. The process of making an appointment was not always clear to the participants; and they did not feel their individual needs (such as sensory impairment and communication requirements) were always taken into account. The complexities of their conditions along with juggling personal factors (for example childcare, work commitments, school timetables) meant that participants sometimes had to balance how many appointments they could go to. Most participants who were offered multiple appointments in one day (at specialist clinics) preferred this, and also preferred additional

appointments where possible to be dealt with at school, over the telephone, or by a visit to the home.

### At the hospital

Difficulties with travel to hospital are exacerbated by sensory impairment, and the rarity of their conditions also meant they often had to travel a long way to specialist facilities.
In looking across different hospitals, some had good lighting and minimised glare, some had good colour schemes which identified areas and furniture, some had clear, good sized signs, some avoided clutter in areas where people were moving, but none of the hospitals managed all of these. Once within the hospital, cafés could be expensive and sometimes they were a long way from the clinic, they were not well lit, and they did not provide for sensory needs. Toilet facilities, obviously important when people are waiting for some time were sometimes easy to access, but others found that sensory issues, made it difficult for them to be independent.

### At the clinic

Patients talked about their experience of reception areas, waiting and the role of specialist co-ordinators. Most frequently they talked about the key needs for face-to-face communication, staff looking while speaking, speaking up and speaking to the patient rather than a companion. Waiting is central to the patients' experience of hospitals. Participants appreciated information about how long they might need to wait, and why, in appropriate formats. Consideration to individual needs and preferences such as appropriate activities for children of different ages, TV with subtitles, or wifi would also be appreciated. Participants worried **a lot** about not hearing their names called for their appointment. Some participants appreciate (or would appreciate) the opportunity to meet other people with their syndrome, but others would prefer this to be through network groups. Clinic co-ordinators, where they existed, performed a vital role as a friendly face, in helping with logistics, and as a point of contact between clinic visits.

## Consultation

In the consultation, participants experienced different levels of communication, with some clinical staff being very aware of individual needs, whilst others were not. Unfortunately, poor communication skills were seen and described far more often than good ones. Participants whose sight was very impaired, were not told what was happening during appointments, such as silence while people wrote notes. Participants were frustrated by staff talking to their companions rather than them and by inadequate or inconsistent provision of communication support (where needed).

Participants needed information which sighted/hearing patients did not, such as being able to touch equipment and they needed information in appropriate text formats (large print, braille, digital). Hearing impaired patients needed spoken information backed up later, such as emailed reports on the clinic visits to read in their own time. Participants also felt strongly that staff should be honest about procedures which hurt or were uncomfortable or inconvenient. Clinics were usually well lit, but sometimes unnecessarily noisy, because doors were left open, or some areas were only bounded by curtains.

## Beyond the clinic

Participants frequently felt they needed some support beyond the appointment, both from the hospital and from other organisations, but they often felt this was lacking. Where there were clinic co-ordinators pathways for support after the clinic were clear, but otherwise they often did not know who to contact. Referrals to support organisations such as Sense would have been welcomed by many, and where they were made, they felt supported. Many participants said they would like to be in touch with others with the same syndrome, and where they were, they found this useful.

## Additional points

Some points about staying in hospital and GP clinics were also raised by participants. Hospital stays were often very difficult for patients with sensory impairments, with hospital staff not understanding their communication and mobility needs. For children and their parents, such

stays were very stressful. In visiting GP surgeries, participants raised again the issues of inaccessible information; and not being able to get consultations in appropriate formats. Where this was working well, they had built up relationships by seeing the same GP over some time.

## Conclusions

For people who had dual sensory impairments and rare syndromes, they found the key areas of mobility, communication and access to information difficult in their hospital visits as in all other areas of life. Their difficulties in each of these areas were in some cases the same as other people's (for example, parking in relation to mobility) but sometimes exacerbated by sensory impairment (for example, signage which they could not see, and they were not able to ask their way because they could not hear the answer) and sometimes was different to that of people who do not have sensory impairments (for example, managing bathroom furniture with no contrast e.g. all white). For each area, some simple strategies would potentially help considerably, for instance, all staff being trained to ask 'Is this communication OK for you?' could allow patients to ask someone to speak up, face them, or read documents to them.

For patients with these rare syndromes, the issues relating to their lives, and their medical treatments all involved connectivity, multiplicity, rarity, and individuality. They needed their services linked together and each aware of what others were doing. They needed clinical staff to understand that their conditions were not just single issues, but each one affected all the others. They knew their conditions were rare and they both appreciated staff who did understand something about it but also those who were prepared to listen to patients.

But, and as an important coda to all the above, these patients were above all, people. They were individuals in the nature of their syndromes and sensory impairments (no two the same) but as well as being patients, participants spoke of being sons and daughters, pupils, musicians, cooks, friends, husbands and wives, mothers, campaigners, sportsmen, artists, and many more identifiers. We would not wish to

forget this rounded perspective in our descriptions of the small element of their lives that was as “patients”.