The impact of Lennox-Gastaut Syndome (LGS) on health related quality of life - a conceptual model

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Introduction

Lennox-Gastaut Syndrome (LGS) is a severe childhood epileptic encephalopathy with onset usually before 8 years of age, and occurrence rates peaking between 3 and 5 years of age (1). LGS is characterised by slow spike-wave electroencephalogram (EEG) disturbance, multiple types of seizures, and intellectual impairment (2). The prevalence of LGS has been reported as 0.26 per 1,000 in a population based study of 10 year old children in Atlanta; LGS accounted for 4% of all childhood epilepsy in this population (3). Tonic seizures are the most characteristic type of seizure in LGS (4) however they are not necessarily present at onset (1). Atypical absences are the second most common type of seizure (4). Other seizures include non-convulsive status epilepticus and myoclonic seizures (1). Seizures are often resistant to therapy (1)

Catastrophic epilepsies that begin during the childhood development stage halt cognitive and social development with long-term effects (5). LGS is considered to be a catastrophic epilepsy because of the poor response to seizures with antiepileptic drugs (AEDs), likelihood of injury from falls related to seizures, and intellectual impairment in >90% of patients (5), all leading to significant psychosocial effects.

The aims of the research were to conduct a formal qualitative research study with care givers of LGS patients to describe the experience of having a child with LGS, and identify issues of concern relating to HRQL of the patient and the care giver. The research aimed to contribute to our understanding of the perceived impact of epilepsy on the HRQL of carers and subsequently to develop two conceptual models: one to illustrate the impact of LGS on a child's HRQL, and one to illustrate the impact on the parent or caregiver's HRQL.

Methods

Semi-structured interviews were conducted with 40 parents of children with LGS in the US, UK, and Italy. This gives Northern and Southern European and North American perspectives of LGS. It was expected that any differences may be due to differences in health care provision and culture. A purposive sampling strategy was applied. Full Institutional Review Board (IRB) approval was obtained prior to commencing the recruitment process. Given the fact that it was not possible to interview the children, parents were asked to provide a report on their perceptions of the HRQL of their child in addition to describing the impact on their own HRQL. Thematic analysis using Atlas.ti was conducted to develop themes relating to the impact on HRQL. The themes were organized into two conceptual models: one to illustrate the impact of LGS on the HRQL of the child, and one to illustrate the impact of LGS on the HRQL of the parent.

Results

Forty parents participated in open-ended interviews. The majority of the participants were female (90%) and white (98%) with a mean age of 43 years. The average age of their child was 14 years and 63% of the children were male.

Impact of LGS on a Child's HRQL

Analysis of the interview data identified several themes related to the impact of LGS on a child's HRQL.

Physical Impact

The seizures are often both frequent and severe, and therefore have a significant impact on several areas of the child's HRQL. Seizures, drop attacks in particular, may cause many injuries for a child with LGS, which would have a detrimental impact on the child's HRQL. This following quote describes some of the injuries suffered by one child:

'If the atonic seizure occurred when he didn't have his protective helmet on, they would always be down the midline of his face. So they would occur around his - the forehead, his nose, his lips, he has lost five of his front teeth, and his chin. And there have been other seizures when he's fallen and he's broken his shoulder, he's broken both shoulders in falls, and there were many cuts and bruises.' (UK)

Cognitive and Behavioural Impact

The child's cognitive impairment may impact on their speech, causing communication problems, their relationships with their peers, the type of recreational activities they enjoy, their ability to care for and feed themselves, and their sleep:

'(Child's name) was normal until she was three, and right now she's 11, and she's probably functioning anywhere between the ages of three and five, so it affects her activities of daily living, as far as how she feeds herself, dresses herself, uses the restroom, things of that nature. Her mobility is affected, her speech is affected ….. pretty much anything that has to do with being a human, she's affected in some way.' (US)

'All these little kids are going, "Well how old is she?" Like, "She's 11." "Why is she in here?" And they're coloring in the lines, and she's dribbling, and it's just bizarre, it really, kind of, puts a barrier between her, and even children that could act like her they just don't understand why she's so large and doing things maybe worse than them, or at the same level as them as, say, a two year old, you know'. (US)

Social Impact

The seizures and cognitive impairment may also impact on the child's social and family relationships, as other children and family members may be more reluctant to spend time with a child who has frequent seizures. Parents described other children not wanting to be friends with their child because of the seizures:

'Since she started seizures nobody has asked for her. Before she used to go to birthday parties, they invited her and she always went, she was welcomed, she had a social life when she was a baby girl, but then nobody has invited her anymore and slowly she has lost all the previous friends'. (Italy)

Treatment

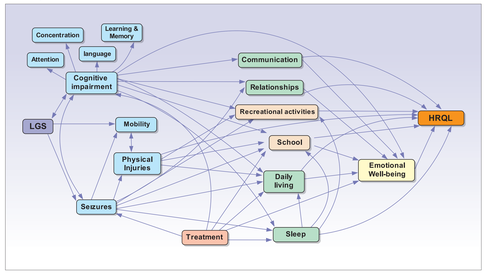
Some parents discussed the positive impact of treatment on seizures. A reduction in the severity or frequency of seizures may improve other areas of the child's HRQL, such as fewer injuries, less disruption at school and less of an impact on their social or family relationships.

'At the worst time, he was on five different anticonvulsants all at the same time, which we thought was a bit radical, but it seemed to help' (UK)

'So, although he's still getting some drop attacks, some tonic clonics and some, you know, longer seizures, the overall improvement, really, I think, has been because of this muzziness isn't so……constant, and that makes his mood better, I think' (UK)

However the parents also mentioned several negative side effects of treatment that they had noticed in their children, such as medication having a 'sedating effect', their child 'seeing funny pictures' or becoming 'really aggressive'.

Conceptual Model: Child with LGS



The arrows in the model indicate the direction of the influence, for example the arrow from school to emotional well-being indicates that experience at school can impact on emotional well-being. The arrows may represent either a positive or negative impact.

Colour Key:
Purple – LGS the condition
Green – Domains of HRQL
Yellow – Psychological impact
Blue – Elements of LGS
Orange – External influences (positive or negative)
Red – Treatment

Impact of Caring for a Child with LGS on a Parent's HRQL

Analysis of the interview data also identified several themes related to the impact of caring for a child with LGS on a parent's HRQL.

Social Impact

Caring for a child with LGS will impact on a parent's social life and family relationships. It may lead to a restricted social life or lead to strained marital relations:

'[Child's dad] and I split up when she was maybe, like, a year old, and he's just - he's not very accepting of [child]. I mean, I think he's, like, really scared and depressed over her, having the condition that she has' (US)

The impact on social life and relationships may lead to feelings of isolation, which may cause depression and impairment in HRQL.

'I got really, really down and depressed, and went through a terrible, terrible stage. And eventually I met some new friends and got out of that depression and I'm really happy now' (UK)

Physical Impact

Sleep may be disrupted due to the child's seizures, which may lead to fatigue, which would also negatively impact the parent's HRQL:

'I suppose I'm completely brain dead because of the lack of sleep and everything'. (UK)

Financial Impact

The data suggest that the parent's career opportunities and finances may be negatively affected by caring for their child, as they may have had to give up their job in order to care for the child. This may lead to anxieties about lack of money:

'I was teaching in a secondary school. I only did six weeks because you just can't cope with… this, you know, that, sort of, a - so I gave up the teaching, and we're in the, sort of, if you like, problem, not, you know, we're not absolutely desperate for money, but we're very worried'. (UK)

Emotional Impact

Some parents discussed a 'sense of loss and unfulfilled expectations', which may lead to the parent feeling depressed, with negative impact on their HRQL:

'It's like the fighting, having to deal with looking at him - I've come to peace with who he is and what he could have been. I let that go a long time ago… So it always comes down to, you know, not letting your heart break every time, being strong, and getting as much rest as possible'. (US)

All parents mentioned feeling anxious; they experienced both anxieties about when the next seizure would occur, and also anxieties about the child's future. The constant fear of seizures caused anxiety:

'The fear when you wake him up and you send him to school, the fear that it will be yet another day where you'll get the phone call, and so it - you are never free of an anxiety - you have to put it to the back of your mind and you have to continue your day. ' (UK)

Parents in each country mentioned experiencing stigma specifically due to their child's seizures rather than the cognitive impairment:

'To protect her for the seizures not for the retardation. I am not afraid if other people see her retardation… only for the seizures… and nothing else' (Italy)

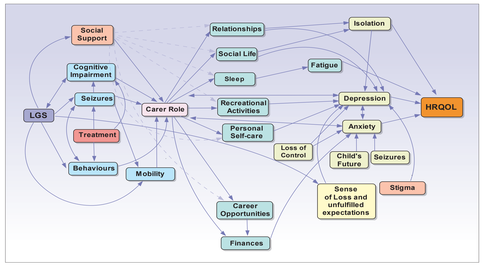
Social Support

Support with caring for their child, either from family, friends or social services, may have a positive impact on parent's relationships, social lives, sleep, recreational activities, their ability to take care of their own health and appearance, and career opportunities. However due to the stigma surrounding epilepsy and cognitive impairments, it can be difficult for parents to find others to care for their child, and therefore a lack of social support may also lead to a decrement in HRQL for the parent.

'I have a lot of emotional support for sure, and I have a good amount of physical support, as far as there is a couple of people in my community that I can trust with taking care of my daughter, if I need them to…… . I don't know how people do it without that, I really don't, because I know that I have it, but there's a lot of people that don't.' (US)

'…a child with LGS, if you were stuck with that 24 /7, I wouldn't be able to cope with it, not at all. I need that break at the weekend, and his dad needs the break in the week.' (UK)

Conceptual Model: Parent of child with LGS



The arrows in the model indicate the direction of the influence, for example the arrow from sleep to fatigue indicates that if sleep is impacted then this may impact fatigue. The arrows may represent either a positive or negative impact. The dotted lines in the model represent the possible positive influence that could come from social support.

Colour Key:
Purple – LGS the condition
Orange – External influences (positive or negative)
Blue – Elements of LGS
Yellow – Psychological impact
Red – Treatment
Green – Domains of HRQL
Pink – Carer role

Conclusions

The qualitative data and conceptual models illustrate the many areas of a child and parent's HRQL that are impacted by LGS and demonstrate the complex relationship between the components of LGS and their impact on HRQL. The models demonstrate the potential areas of HRQL that may be enhanced for both the child and caregiver if the symptoms of LGS were improved.

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