

Parents' Experiences Of Having A Child Diagnosed With Landau Kleffner  
Syndrome

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## ABSTRACT

**Background and Aims:** Landau Kleffner Syndrome (LKS) is a rare childhood neurological disorder that is characterised by epileptic disturbance and acquired language regression. The current literature on LKS takes a predominantly medical stance, with little attention given to the affect this syndrome has on the family. Through the adoption of a qualitative design, the current, United Kingdom based, study aims to explore parents' subjective experiences of having a child with LKS and how they cope.

**Method and Results:** Eight interviews were conducted with parents whose children had been previously diagnosed with LKS. Each interview was transcribed and thematic analysis conducted on the data. Two themes were identified, each indicating important aspects of the parents' experience of LKS: 1) Challenges to coping 2) Evolution of the family roles and ways of coping over time.

**Discussion:** Findings highlighted challenges throughout the course of LKS. At the initial onset, parents particularly emphasised the significance of loss, while witnessing the deterioration of their child's behaviour, skills and health. This was further exacerbated by the rarity of the disorder, which made accessing helpful formal and informal support challenging, because others typically held little knowledge about LKS. Parents reported a number of consequential changes to their lives including parenting style, managing finances, social interaction and a need to advocate. The range of ways parents coped included the acknowledgment of positive experiences, drawing from personal resources and taking one day at a time. Based on these findings, clinical, service and research level implications are considered.

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## **1. CHAPTER ONE: INTRODUCTION**

Landau Kleffner Syndrome (LKS) is a rare childhood neurological disorder that can have a widespread functional impact on a child's quality of life. Largely managed in medical settings, the experience of parents<sup>1</sup> of children with LKS has attracted little research attention.

This chapter begins with a definition of LKS, outlining the specific clinical features. Next there is discussion of the broad theoretical contexts which have led parents to be prioritised within policies, clinical practice and research, followed by a consideration of the research which has begun to explore the parental experience of LKS. In the context of the minimal amount of research available, an explanation is provided of the necessity to draw from more widely researched disorders, focussing on key overlapping clinical presentations, with the aim of creating a greater understanding of how parents may experience LKS. The chapter ends with a rationale for why further research is needed, and outlines the current study's aim to explore parents' experiences of having a child diagnosed with LKS.

### **1.1. Literature Review**

An electronic literature review was initially conducted to explore families' experiences of LKS. As little literature was available, a wider search was conducted to include parental experiences of other childhood health disorders with relevant, overlapping clinical presentations. Literature was identified within the databases EBSCOhost, PsychARTICLES, PsychINFO and Science Direct. Appendix A lists the terms and parameters used to search for relevant material. Further to these searches, a "snowballing" technique was employed to highlight additional references which may not have been otherwise captured.

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<sup>1</sup> The term Parent will be used to describe an individual who is the primary caregiver of a child.

## 1.2. Landau Kleffner Syndrome (LKS)

### 1.2.1. Features and Onset of LKS

Described as an epileptic encephalopathy, LKS is characterised by epileptiform electroencephalographic (EEG) abnormalities during sleep and acquired aphasia (Caraballo, Cejas, Chamorro, Kaltenmeier, Fortini, & Soprano, 2014). The presence of epileptic seizures is not a prerequisite for LKS and their occurrence varies greatly within this disorder (Caraballo et al., 2014). Partial and generalised seizures have been observed (Fandino, Connolly, Usher, Palm, & Kozak, 2011) as well as an absence of seizures in 20% to 30% of the affected population (Caraballo et al., 2014). Remit

Symptoms of LKS typically begin between the ages of three and seven, and then stop after adolescence. Males are affected more frequently than females (ratio 1.7:1) (Behr, Goltzene, Kosmalski, Hirsch, & Ryvlin, 2016; Caraballo et al., 2014). The key clinical feature of LKS is the occurrence of acquired aphasia, after a period of typical language development. Presenting initially as a verbal auditory agnosia, a global deterioration of language abilities is subsequently observed (Fernández, Loddenkemper, Peters, & Kothare, 2012). The child initially loses the ability to give semantic meaning to different sounds and therefore to comprehend language. This is followed by expressive aphasia, where there is a reduction in spontaneous speech. In some cases, these difficulties with language communication can extend into mutism (Caraballo et al., 2014). This deterioration of previously intact language skills can be a gradual or sudden loss (Fandino et al., 2011). Clinical professionals typically refer to this presentation as the 'active phase' of LKS (Downes et al., 2015).

Although research highlights the need for further investigation into the neuropsychological dimensions of the condition, non-verbal skills are understood to be sustained in most children with LKS (Titus, 2017). Intelligence measures which predominantly utilise non-verbal tasks (including direction and responses) have found children to be performing in the 'average' range (Van Bogaert et al., 2017; Titus, 2017). Consequently, the relative strengths observed in skills such as visual processing, allow the child to continue communicating if appropriate

adaptations are made. For example, the use of sign language, communication boards and specialist computer programs have proved beneficial (Cockerell, Bølling, & Nakken, 2011).

### 1.2.2. Behavioural, Functional and Mental Health Sequelae

Regardless of the degree of language deficit, dramatic behavioural changes are commonly reported within the active phase of LKS. Although they are not acknowledged to be a key clinical feature, reports frequently highlight increased inattention, hyper-activity, oppositional behaviour, aggressiveness, mood variability and some psychotic experiences (Caraballo et al., 2014; Cockerell et al., 2011; Malvestio, 2010).

Consequently, the clinical features of LKS can overlap with more frequently occurring difficulties such as hearing impairments, a learning disability, or emotional and behavioural difficulties (Malvestio, 2010; Fandino et al., 2011; Pullens, Pullens, Blau, Sorger, Jansma, & Goebel, 2015). The risk of a misdiagnosis of an Autism Spectrum Disorder (ASD) can also occur, as both disorders present a deterioration of previously acquired cognitive and social skills (Malvestio, 2010; Fandino et al., 2011). Fandino et al. (2011) outline the importance of professionals being aware of the defining behavioural features of ASD, highlighting that a child with LKS will still have the desire to communicate with others.

This is particularly pertinent in the context of many children also having intact non-verbal intelligence. Explanations for the behaviour changes observed can therefore be understood as a result of both the epileptic disturbance and the frustration associated with having profound difficulty in communicating (Malvestio, 2010; Nieuwenhuis & Nicolai, 2006).

### 1.2.3. Course and Recovery

The symptoms of LKS move between periods of stabilisation and deterioration over months or years, before typically stopping in adolescents. For unclear reasons, the epileptiform activity tends to remit after adolescence and neurocognitive recovery often occurs (Caraballo et al., 2014; Malvestio, 2010).

This signals the end of the active phase. However, there is a lack of homogeneity seen within the developmental trajectory of LKS and some children recover their language abilities whilst others experience lifelong difficulties (Metz-Lutz & Filippini, 2006; Caraballo et al., 2014).

A dominant theme highlighted by professionals, is the challenge to ensure an early and accurate diagnosis. Behaviours suggestive of epileptic seizures are sometimes not present and initial EEGs can show complex and sometimes inconclusive findings (Caraballo et al., 2014; Fandino et al., 2011). The clinical features of a verbal auditory agnosia can also be difficult to identify at the early stages, as a subtle deterioration of ability may be disguised by the child's ability to lip-read or follow suggestive cues from their environment (Fandino et al., 2011).

#### 1.2.4. Etiology

As with many types of epileptic syndromes, the etiology of LKS is still unknown and there is no dominant explanation for how the EEG abnormalities create the presentation commonly observed. Although a cerebral pathology is understood to cause cerebral dysfunction, the impact is more commonly seen to cause permanent cognitive difficulties. The fluctuation in the functioning seen within children with LKS is therefore not easily explained (Titus, 2017). Furthermore, research indicating that children with LKS experience a range of subtle differences in their language presentation, suggests that there may also be differences in the etiology for different children (Cockerell et al., 2011).

Despite this unclear etiology, a number of recent studies have highlighted the role of genetic mutations, specifically with GRIN2A (Pierson, et al., 2014). However, this does not account for all cases of LKS, and environmental influences cannot be excluded as the sole cause or as a modifier of a genetic cause of LKS (Caraballo et al., 2014; Conroy et al., 2014).

#### 1.2.5. Prevalence and Risk

LKS is recognised as a rare syndrome (National Institutes of Health (NIH), 2015), with Kaga, Inagaki and Ohta (2014) calculating the incidence (978,000) of children aged five to fourteen with LKS to be one in a million. Despite there being over 400 studies referencing LKS, the study within Japan by Kaga et al. (2014) presents the only systematic epidemiological investigation (Behr et al., 2016). Kaga et al. (2014) suggest that few epistemological studies are undertaken because the clinical signs and symptoms of LKS are not well known to paediatricians, and their seizure types and characteristics are not specific to LKS.

#### 1.2.6. Treatment

As the etiology and developmental trajectory of recovery is still being investigated, clinicians are also faced with a challenge in selecting the best treatment management pathway to reduce neurological damage whilst children are in the active phase. A delay to a child receiving an accurate diagnosis and appropriate treatment are understood to significantly affect the cognitive and behavioural prognosis (Besag, 2006).

Guidelines for treating LKS suggest that pharmacological interventions must be given early and aggressively to avoid language deterioration (Caraballo et al., 2014). Antiepileptic drugs (AEDs) such as valproate, ethosuximide, clonazepam, or clobazam have been demonstrated to be effective. Other AEDs are understood to be avoided because they can exacerbate the damaging epileptiform activity (Caraballo et al., 2014). Corticosteroids have been reported to improve outcomes (Sinclair & Snyder, 2005). Surgical procedures to control the epileptiform activity have also been used, however more evidence is needed before prioritising this intervention over alternatives (Downes et al., 2015).

### **1.3. Understanding the Impact Childhood Health Disorders have on the Family and how they Cope**

Between 10% and 30% of children are affected by chronic illness or physical health problems (Eiser, 1995 p.27). Many children and their families find this experience extremely challenging, with research presenting an increased risk of

psychosocial and emotional difficulties (Waldboth, Patch, Mahrer-Imhof, & Metcalfe, 2016). To understand this relationship and its relevance to current paediatric research, it is important to consider the theoretical context that shapes our knowledge-base, policies and clinical practice.

### 1.3.1. Recognising the Importance of Family in Supporting the Effective Treatment of Children

Over the last 60 years, our understanding of the impact childhood disorders have on children and the people around them, has shifted. Moving away from a 'dominant expert' model, where the professional determines the needs of the client, society now prioritises the experience of the child and family (Falvo, 2013). This change is reflected in major policy changes, the research conducted and the ways in which health-care and related services are developed and delivered to children with chronic illnesses.

The change to visiting policies within children's hospitals is an illustrative example. Up until the 1950s restrictive visitations were still in place, with parents advised not to see their hospitalised children due to the dominant belief that parents would serve as a barrier to effective care, in part because the child would become distressed when the parents left (Junge, 1987; Shields, Pratt, Davis, & Hunter, 2007).

In response to parents of children with childhood disorders advocating for increased involvement in their child's health and related care, researchers and policy makers began to explore the importance of the family (MacKean, Thurston, & Scott, 2005). The seminal research by Bowlby (1958) and the influential recommendations within the Platt Report (Central Health Services, 1959) saw this enforced separation as having detrimental effects on the emotional, psychological and physical wellbeing of both the child and their family. Consequently, the British government recommended unrestricted visiting, the encouragement of mothers to stay with their child, and the prioritisation, by hospital staff, of the emotional needs of children (Shields et al., 2007). Parents were now formally recognised as active participants in their child's care, and no longer positioned as a burden to be ignored.

### 1.3.2. Understanding the Impact of Childhood Health Disorders: The Underpinning Influential Frameworks of Knowledge

Enhancing the quality of children's lives is now a national and international priority. The impact of childhood disorders has been explored through the synthesis of knowledge across many domains. This section aims to summarise the influential thinking that has shaped current societal and professional understanding of the impact childhood health disorders have on the family.

#### *1.3.2.1. Normative Models of Child Development*

Developmental theories present childhood as a critical period within the lifespan. Key theorists such as Erickson (1956), Bowlby (1978), Piaget (1952) and Vygotsky (1978) characterise young children's level of functioning as a progression through distinctive phases and/or milestones. Children's development is now understood to be a complex interplay between the biological, the cognitive and the social skills and capacities they develop. The relationship to key attachment figures was identified as crucial, not only for physical survival but for emotional security, social integration and the development of cultural competencies (Woodhead, 2006).

This knowledge has consequently provided a framework to navigate typical development, promoting awareness of the 'protective' and 'risk' factors that affect children's long-term well-being. The importance of children accessing adequate resources and interacting with caring and attentive care-givers are key themes within this literature (McDonald, Kehler, Bayrampour, Fraser-Lee, & Tough, 2016). 'Risk' factors highlight the impact of not having access to these protective factors and experiencing individual 'vulnerabilities', such as poor childhood health (McDonald et al., 2016; Woodhead, 2006).

In the context of childhood health, these frameworks create societal and thus parental expectations of children's behaviour. Unmet milestones are often the first indicators, for parents and professionals, that the child is not developing typically (Harden, Black, & Chin, 2016). Whether the result of biological or

environmental factors, the speed and content of interventions are also shaped by child development theories that present early support (formal or informal) and continued acquisition of adaptive skills as crucial factors in reaching important milestones and improving outcomes in later life (Kover, Edmunds, & Weismer, 2016).

#### *1.3.2.2. Emotional Impact of Unmet Normative Milestones*

The literature exploring parents' experiences of their children's unmet milestones, as the result of childhood health disorders, frequently highlights themes of loss and grief (Smith, Cheater, & Bekker, 2015). Drawing on the commonality between the parental responses to the loss of a child's abilities and the loss of a child through death, bereavement grief models (Kubler-Ross, 1969; Parkes, 1986; Strobe & Shut, 1999) have often been applied to conceptualise the experience for parents whose children are diagnosed with a childhood disorder (Anderegg, Vergason, & Smith, 1992). This is understood by some as grieving the metaphorical 'death' of the idealised child (Hewson, 1997).

#### *1.3.2.3. Systemic Perspective*

A systemic perspective has been utilised within the literature, as it considers the broad range of factors that impact the family's experience of childhood disorders (Cohen, 1999; Cipolletta, Marchesin, & Benini, 2015; Fredman, 1997, p.1; Kazak & Nachman, 1991; Masarik & Conger, 2017). Acknowledging the variations in family responses to illness, this theoretical approach looks beyond a sole focus on the individual, and values the influence of family, societal and cultural context.

The Family Life Cycle (Carter & McGoldrick, 1999) is an example of a systemic model where family context is explicitly considered. Through the exploration of multiple positions, interdependence is assumed within the family, i.e. what happens to one family member affects other family members. It also highlights the influence of values and beliefs on the interaction and function of the families.

From this perspective, the variation of parental experience is considered a consequence of the particularities in which illness is perceived and managed. Consequently, it is not assumed that the childhood disorder itself causes distress,

but rather the consequential social, psychological and physical challenges that arise when transitioning across the expected stages of family life (DePape & Lindsay 2015; Waldboth et al., 2016). Parents are presented with an unexpected developmental trajectory for their child, which may bring about complexities to their immediate and future parenting roles (Fletcher et al., 2016).

The social-ecological perspective (Bronfenbrenner, 1979) goes further to present a systemic framework which allows a rich and dynamic picture of societal and cultural influences. Prominent research into families coping with childhood health disorders frequently utilise this model to identify the ecological systems around a child, embedding childhood experiences within the context of micro- to macro-level influences (Kazak & Nachman, 1991). Conceptualised as concentric circles around the child, cultural values, beliefs and norms are positioned furthest away from the child, whilst people and institutions are placed nearest, representing a more direct influence. Each are thought to have a bi-directional influence on the child and family.

For example, in the work of Kazak and Nachman (1991) they consider the impact societal and cultural context has on the parent's experience and highlight that many factors come into play. Access to different communities (e.g. places of worship, workplace, schools and hospitals) develops one's framework of knowledge. In the context of childhood disorders, they highlight that this knowledge affects our understanding of the child's disorder aetiology, symptoms and prognosis.

When considering the impact of western culture, importance is often placed on social independence and economic success. These values can influence the expected stages of family life and parental roles, and underpin the common worries about a child's ability to live independently and be financially secure (Waldboth et al., 2016). On a wider level, these values influence a societal discourse around 'societal worth', framing childhood interventions as a means to "produce a healthy adult population that is sufficiently skilled to participate effectively in a global economy..." (Shonkoff et al., 2012, p. 232).

The repercussions of prioritising 'societal worth', are the marginalisation of groups of people due to their abilities and attributes not being valued when held up against socially constructed norms (Goffman, 1963). The experience of stigma is widely reported within families of children with childhood disorders and can be framed within the neoliberal historical context (Francis, 2012). Medicalisation and problematisation of childhood leads to constructions of parenthood which place parents as solely responsible for their child's difficulties. Consequently, parents may experience guilt and shame which can initiate or exacerbate isolation or exclusion from peer groups and communities.

#### *1.3.2.4. Paediatric Models: A Biopsychosocial Perspective*

There has been an increasing recognition that paediatric care from psychosocial professionals needs to acknowledge the diverse range of factors that affect a child's quality of life. Current paediatric practice has moved beyond a narrow focus on health, but strives to promote psychological and emotional wellbeing of the child and family.

The Biopsychosocial model is an example of this position. Paediatric professionals are encouraged to embrace a holistic perspective that includes the biological aspects of the health experience (e.g. pain, cognitive ability, prognosis, and visibility), psychological implications (e.g. stress, grief) and sociological factors (e.g. family support). In the work of Cohen (1999), this model is drawn from to emphasise how the effects of a health condition are dependant on the particularities of each of these factors and consequently influences how a families respond and cope. I(

This framework continues to be hugely influential, and has been utilised by the World Health Organisation (WHO, 2007) to create a universal categorisation system: International Classification of Functioning, Disability and Health for Children and Youth (ICF-CY). The ICF-CY aims to provided a conceptual framework for professionals and family members to define development, functioning and health (WHO, 2007). Focusing on the dynamic interaction between disability and function, this sophisticated categorisation system

highlights health and wellness within the context of an individual's social and environmental context. This importantly includes the interaction with family and other close caregivers. As a consequence, assessments and interventions can incorporate this holistic perspective and be developed and delivered accordingly.

As seen within the social model of disability (Abberley, 1987), a deficit model is neglected in favour of the positive attributes within one's life being valued and prioritised. The value of the family is evident within this framework and the resources and strength that the family give the child, and the child brings to the family, can readily be acknowledged.

### 1.3.3. Understanding the ways in which Families Cope

The literature has moved beyond a sole focus on the challenges experienced by the families who encounter a childhood disorder and now incorporates research into how they cope (Atkin & Wager, 2000). Coping is understood to be a complex phenomenon, with theories drawing from individual, family and societal factors (Choen, 1999; Kazak & Nachman, 1991; Knafel & Deatrick, 1986).

#### 1.3.3.1. *Stage Models: an individual perspective*

Some theorists have conceptualised the parents experience of childhood disorders into a process of stages where the parents' adaption and ability to cope are dependent on the resolution of their loss; this is achieved through the 'acceptance' of their child.

Researchers have given many names to these stages of adaption and coping (Hewson, 1997). However, the work of Sen and Yurtsever (2007) presents an interesting and comprehensive account of parents' general reactions, with three main categories outlined: primary reactions, secondary reactions and tertiary reactions.

Primary reactions focus on the initial shock and suffering experienced by parents when they become aware of their child's difficulties (Sen & Yurtsever, 2007). These reactions have been repeatedly noted since an academic interest in this

area began (Fletcher, Flood, & Hare, 2016; Smith et al., 2015). Usually focusing on the context of diagnosis, literature has highlighted two areas of interest: the devastation of hearing unexpectedly from a professional that something may be wrong with their child's health (Harnett, Tierney & Guerin, 2009) and the parental distress of experiencing professionals' resistance to acknowledging their anxieties about the child's atypical behaviour or development (Fletcher et al., 2016).

However, literature also acknowledges that receiving a diagnosis may engender positive emotions, due to parents experiencing validity for the concerns they have been expressing, or being able to access appropriate support and interventions (Ho, 2004; Lauchlan & Boyle, 2007).

Sen and Yurtsever (2007) also place denial as a co-occurring primary response. Described as a defence mechanism activated through fear of facing an unknown, it is thought to be an essential prerequisite to acceptance and coping. If not prolonged, this adaptive strategy is thought to allow parents to protect themselves from the overwhelming emotional response and focus on the positives of their situation (Kearney & Griffin, 2001). The presentation of the child's difficulties is noted to affect the parent's ability to reach 'acceptance'. For example, an uneven cognitive profile can cause confusion and lead to difficulties in understanding the severity of a child's condition (Fletcher et al., 2016).

The work of Knafl and Deatrick (1986) go on to describe the potential role of normalisation, linking its function to that of denial. Whilst acknowledging the existence of an impairment, the parent is thought to place value on presenting and engaging in behaviours which promote their family life as normal and the social consequences of the impairments as minimal.

Sen and Yurtsever (2007) go on to describe feelings of guilt, indecision, anger and shame as secondary responses. These emotions are widely reported within the literature (Fletcher et al., 2016; Smith et al., 2015). In response to the grief experienced primarily, parents are understood to question their actions and the control they might have had over the outcome of events (Ellis, 1989).

Consequently, anger and blame can turn inward, making it challenging to feel happy (Findler, Jacoby, & Gabis, 2016).

The tertiary reactions are presented as an ongoing phase, where parents bargain for help from professionals and accept that changes need to be made in their lives (Sen & Yurtsever, 2007). The parent begins to develop an understanding of their new, unexpected situation and the strong emotions outlined in the previous stages subside (Smith et al., 2015). Parents become familiar with a typically unknown world of medical and psychological information, and develop familiarity with the systems in place to support their child (Russell, 2003).

The challenge in applying a grief model within the context of parental experiences, mirrors the critiques expressed within the bereavement literature (Hewson, 1997). The rigidity of their application can lead to unhelpful assumptions which simplify the complexity and diversity of experience. Although Sen and Yurtsever (2007) propose that their stages can vary in length and intensity, and be experienced in a non-linear fashion, there is an assumption that feelings of grief are eventually resolved and that this resolution is intertwined with acceptance of the child's difficulties and viewed as a final position.

Researchers opposed to a stage model framework argue that resolution is a continual process which is related to ongoing, significant family life-cycle transitions. A conflict between the parents' unconscious desires and expectations of their child, and the child's actual development, can reintroduce feelings of loss and grief at each developmental milestone. This repeated experience has been described as 'chronic-sorrow' (Copley & Bodensteiner, 1987). Rather than a continuous state, it is acknowledged that the parent will encounter episodic moments of distress alongside positive feelings (Hewson, 1997).

Other critiques highlight the importance of acknowledging the individuality of parents' experiences of coping. Parents present varied responses when encountering the loss of their child's abilities and this does not always fit within structured stages (Hewson, 1997). Parents' adjustment and ability to cope is instead thought to be dynamic, as their child's condition and family-needs

change. Responses to these changes are met with different degrees of stress, relating to the personal meaning of the loss, the wider context this occurs within and the resources perceived to be available (Hewson, 1997).

#### *1.3.3.1. Coping Strategies: A Family Perspective*

A strong theme within the current coping literature, is that a predominantly deficit orientation which assumes only a psychopathological reaction to the experience of stress is unhelpful. Instead, exploring what it means for a family to cope 'normally' with an abnormal event, is critical in allowing the challenges faced to be considered alongside the strengths and resources used to promote resilience and coping (Atkin & Wagar, 2000; Rolland & Walsh, 2006; Knafl, Deatrck, Knafl, Gallo, Grey, & Dixon, 2013; Kazak & Nachman, 1991; Cipolletta et al., 2015).

Cohen's (1999) review on research exploring families coping with childhood chronic illness highlights the positive impact resources within the family system can have on illness management. Exemplified through the description of 'balanced coping', Cohen (1999) details the positive affect family's can have when they are able to flexibly respond to the needs of the illness, family and self (Patterson et al. 1993).

Atkin and Wagar (2000) go on to explore the role of 'balance' and suggest that 'boundary setting' also aligns with this parental position. This coping strategy highlights the benefits of developing an element of separation from the role of caring and maintain a value for having some autonomy. This position helps avoid 'engulfment', where the parent's carer role becomes the centre of their identity and they find it challenging to separate themselves from the child's suffering.

These strategies are thought to be a repercussion of the meanings and meta-values created from the family systems past experience or understanding of loss, illness and caretaking (Cohen, 1999). Kazak and Nachman (1991) also values this family systems perspective, and highlights that a family's origin or pre-illness functioning may be critical in influencing which coping strategy is implemented. Kazak and Nachman (1991) exemplifies this through the description of 'problem focused' and 'emotion focused' coping seen within Hughes, and Chesler's (1985) research. Claiming neither one to have more helpful outcomes, the research

highlights that different families and parents utilised different strategies based on their own frame of reference.

#### *1.3.3.2. Resilience Models: A Ecological- Systems Perspective*

Whilst acknowledging that families are devastated by chronic stress from a variety of situations, literature has begun to focus on the experience of increased strength and resourcefulness which enables families to positively adapt (Walsh, 2003). Within paediatric psychology literature, the concept of resiliency is becoming more dominant. Mullins et al. (2015) highlights that the important commonality across resilience models, is their framing within wider systemic contexts. Best described in the work of Bronfenbrenner (1979), parental coping is understood to be the interplay between numerous child, parent, societal and cultural variables.

For example, the risk-resistance adaption model (Wallander et al., 1989) hypothesises that psychosocial adjustment is positively related to resistance factors (e.g., family cohesion, adaptive coping styles) and negatively related to risk factors (e.g., poverty and lack of social support).

Thompson and Gustafson's Transactional Stress and Coping Model (1996) model, expands on this relationship and suggests that adjustment is further impacted by illness-specific variables (e.g., disease type, diagnosis, and illness severity), demographic variables (e.g., socioeconomic status, gender, and age), and various intrapersonal adjustment processes. Within this model, emphasis is also placed on the interactions between child and parent adjustment. Described as a reciprocal influence, child adjustment is thought to influence parent adjustment, and in turn, parent adjustment influences child adjustment).

Kazak and Nachman (1991) social ecological model explicitly names the many systems (i.e., child, family, social group, school, community, and culture) in which the child and parent live and experience the childhood disorder. Drawing attention to the interactive nature between the child's diagnosis and each ecological system. For example, Danseco (1997) and Atkin and Wagar (2000)

found that some parents sought comfort and reassurance by prioritising a spiritual understanding over the dominant biomedical explanation.

Furthermore, parents are seen not to be passive receivers of experiences. Instead meaning-making (e.g. around childhood health) within families and others in the network, is influenced by wider societal beliefs. This is explicitly addressed within the Coordinated Management of Meaning (CMM) model (Cronen, Pearce, & Changsheng, 1980). Focusing on the bi-directional nature of hierarchical levels of context, CMM is a systemic model which argues that individuals are not passive receivers of their social world, but that changes to one level of context can have an effect on others. Therefore, not only can individuals or families be understood to be influenced by these downward 'contextual forces', but they can also respond, challenge and work towards changing pre-existing meanings through upward 'implicative forces'.

In the context of childhood health, the construction and communication of meaning between families and other levels of context can be seen to have gradually shaped society's wider understanding. Drawing from the previous example mentioned, the change to parental visitation rights within hospital (MacKean et al., 2004) and the shift away from a dominant expert model can be understood as the outcome of the gradual changes to meaning which have shifted society into prioritising the experience of the child and family (Falvo, 2013).

#### **1.4. Research, Policy and Clinical Practice: Locating the Child Within their Social and Family Context**

In the current context of child development and childhood health literature, emphasis is placed on locating the child within their social and family context. The importance of psychosocial support for families and for children with chronic illness or disability, is recognised within government guidelines such as the National Service Framework for Children in Hospital (DoH, 2004) and Making Every Young Person with Diabetes Matter (DoH, 2007). Both highlight the

importance of professionals within paediatric settings to address the needs of the child and family. This position promotes parents becoming partners in decision-making and developing an expert role. In turn this leads to more informed and active involvement. A number of names and frameworks have been developed to formalise this way of working. These include; family-centred care (Feeg, Paraszczuk, Çavuşoğlu, Shields, Pars, & Al Mamun, 2016; partnership-in-care and parental involvement (Shields et al., 2007).

Research and policy are not thought to relate in a linear way; instead they are best understood as functioning in parallel. Both shape, and are shaped by, the broader cultural, political and economic context (Woodhead, 2006). Batalden et al. (2015) highlight the financial drive to formalise the prioritisation of the family within research and practice. The government's push for the co-production of public services, partly through the use of service-user involvement, aims to create resource-efficient services. By responding directly to the user's need, waste and cost is assumed to reduce.

As with other government cost-cutting initiatives, the political motivation to promote service-user and family involvement has been viewed with some suspicion. Cowden and Singh (2007), suggest that it can create a false sense of power equality. Under a guise of promoting service development and expansion, it can in fact become a process where agendas are set around how to utilise existing or diminishing resources. Cowden and Singh (2007) consequently urge that collaboration with service-users and families be authentic and aspire to contribute to a process of creative and critical dialogue with professionals.

### **1.5. Implications for LKS literature**

Emphasising the importance of the family within paediatric literature has led to an acknowledgment of their experiences and has more readily presented parents as a potential source of support and knowledge. Their contributions not only have the potential to positively affect the child's health and wellbeing outcomes directly, through increased attentive caregiving (Feeg et al. 2016; Brilli et al.,

2014; Kuhlthau et al., 2011), but they have also shaped the development, delivery and evaluation of services.

As a consequence, paediatric researchers have continued to present the importance of involving the family by making them the focus and participants of their studies. Although siblings' experiences are still relatively under researched, the particulars of parental experiences across different childhood health disorders are becoming more frequently explored (Eiser, 1995, p.175; Edwards & Titman, 2010, p. 55).

LKS research has begun to follow this trend and an exploration into the specific biopsychosocial issues of this childhood disorder has begun to build momentum. Although the current research is dominated by a medical perspective and predominantly brings attention to the experience of professionals within paediatric settings, a few studies have explored parents' experience of LKS (Cockerell et al., 2011; Lemard-Reid, 2014; Wairungu, 2015). With the aim of developing an understanding of this rare health disorder, exploration into the parents' experience can be understood as a crucial development in LKS research.

## **1.6. Parental Experience of LKS and their ways of Coping**

This section aims to review the small amount of literature pertinent to the parental experience of having a child with LKS. Through the identification of key themes within studies which have explored parents' experience of LKS, a clearer understanding of the specific biopsychosocial features of LKS can be developed. This review also highlights gaps of knowledge within the literature, presenting where there may be opportunities to draw from literature on more widely investigated disorders and where further research could help to gain a deeper insight into parents' experiences of LKS.

### **1.6.1. Process of Gaining a Diagnosis**

A retrospective study by Cockerell et al. (2011) presents information gathered from both children's medical records and semi-structured interviews. The

research aimed to describe the developmental trajectory of LKS and explore the parents' experience of accessing the health and educational systems within Norway. Parents were initially asked about the diagnostic process and consequence of their child receiving a diagnosis of LKS. Most described a challenging process, where the journey to establishing an accurate diagnosis had led them to contact, on average, three different authorities. Parents often felt that their concerns were not taken seriously and highlighted the challenge of professionals' lack of awareness of the syndrome.

Misdiagnosis of LKS was common to two studies, with parents whose child presented with a difficulty in auditory discrimination, often being given an inaccurate initial diagnosis of a neurodevelopmental disorder (Lemard-Reid, 2014; Cockerell et al., 2011). Lemard-Reid's (2014) doctoral thesis highlights more specifically the opportunities where parents could act to prevent LKS misdiagnosis. Conducted within the United States of America (USA), the study describes parents' initial concerns arising when milestone regression, epileptic symptoms and verbal and auditory difficulties are observed. Evaluations were then typically carried out by the child's paediatrician or emergency department. When subsequent treatments did not improve their child's symptoms, Lemard-Reid (2014) reported that parents became increasingly concerned about their child's academic performance, behavioural presentation and language abnormalities. In turn, parents became sceptical about the initial diagnosis given and sought further explanations. Both Lemard-Reid (2014) and Cockerell et al. (2011) identified the difficulty in communicating with health care providers as a significant challenge to gaining the definitive diagnosis of LKS. In response, parents described feeling alone, helpless, and overwhelmed (Lemard-Reid, 2014).

When parents did receive a definitive diagnosis of LKS, most were satisfied with how the diagnosis was communicated and the subsequent information, guidance and emotional support they received (Cockerell et al., 2011). Although initial responses were mixed, with some parents shocked and others relieved (Cockerell et al., 2011), it was acknowledged that the diagnosis helpfully allowed for a better understanding of the child's difficulties and ensured more appropriate

help in school (Cockerell et al., 2011; Lemard-Reid, 2014).

#### *1.6.1.1. Parents as Advocates and Unintended Inequalities*

During this journey to gaining a diagnosis of LKS, Cockerell et al (2011) and Lemard-Reid (2014) also highlight the parental belief that it is a necessity to undertake an advocacy role. By using their knowledge and financial resources, parents were able to fight against an incorrect diagnosis or access alternative treatment and resources. Similar to findings in other studies (Berkman, Sheridan, Donahue, Halpern, & Crotty, 2011), Lemard-Reid (2014) identified that children of parents with higher education and increased health-related knowledge were able to make decisions for their children that positively influenced speed of diagnosis and treatment. Furthermore, ethnic background was understood to compound socioeconomic barriers to a LKS diagnosis, as parents perceived themselves to be viewed as less credible with professionals. Some parents reported instances of their children's symptoms being viewed as the consequence of their parenting ability (Lemard-Reid, 2014).

#### 1.6.2. Challenges Experienced by Parents and Teachers

Wairungu (2015) completed a thesis which utilised qualitative methods to explore the parental experience of LKS within the USA. This research specifically investigated experiences related to teaching and 'serving' an individual diagnosed with LKS. Through a single case study, the researcher interviewed teachers, the parents, and collaborating personnel who had taught and interacted with a particular child named Grace. This research identified four prominent challenges: speech and language, behavioural, economic resources and social skills. Although Wairungu's (2015) study predominantly focused on the professional experience, I aim to prioritise the parental reports to gain add further insight into the parental experience.

The difficulty in processing or expressing spoken language was one of the most commonly mentioned challenges. Grace's parents described her decline in ability as significant and observed that she would often become agitated when she was unable to communicate with others effectively. This agitation would frequently precede behaviour experienced as challenging. Behavioural challenges were

described as verbal and physical aggression.

The theme of economic challenges encompassed the expenses reported at both home and at school. School noted the necessity to provide a highly qualified teacher, a teacher's assistant and relevant crisis training for all their staff. Parents highlighted the money spent on replacing destroyed furniture and technology, as well as the expense of gaining a diagnosis and medication. This theme is mirrored within Lemard-Reid's (2014) research, which identifies the financial expenses related to the specialist evaluations.

Parental reports within Wairungu's (2015) study also gave many examples of Grace finding it challenging to engage with others socially. These examples included intruding on others' personal space, sharing personal details in public and asking inappropriate questions. Consequently, Grace's teachers and parents shared concerns about the challenges she may experience in her life post-school and worried that Grace's difficulty in developing friendships would make it hard for her to interact more independently within the wider community. Finding a service-provider who would be able to meet Grace's varying needs was also a concern raised by her parents.

### 1.6.3. Valued Resources and Interventions/Approaches to Cope with Challenges

A number of resources and interventions were identified as valuable in coping with the presenting challenges. Cockerell et al. (2011) assert that professionals were highly valued for their support in addressing the child's behavioural and educational needs. More specifically, the paediatrician and the speech and language therapists within the national epilepsy service were considered the most important professionals when the diagnosis was provided, whilst local teachers and speech and language therapists were identified as key figures post-diagnosis.

Lemard-Reid (2014) and Wairungu (2015) identified beneficial practical strategies. These strategies included the use of behaviour intervention plans, where the importance of accurate data collection to correctly identify antecedent events, triggers, and consequences of behaviour was emphasised. The use of

visual aids was advised to support understanding and effective communication. The repetition of information was also understood to be important, as it allowed the child increased opportunity for comprehension and learning.

Lastly, all three studies highlighted the importance of parents having a strong support system of co-workers and family members. In the context of family-professional relationships, parents valued professionals who had the ability to draw from flexible and creative intervention approaches with their child (Cockerell et al., 2011; Lemard-Reid, 2014; Wairungu, 2015). Communication between the national and local services was understood to be a great asset in promoting this (Cockerell et al., 2011).

### **1.7. Parallels Between LKS and Other Health Presentations: Clinical Features Which Impact The Families Experience And Subsequent Coping**

The studies conducted by Cockerell et al. (2011), Lemard-Reid (2014) and Wairungu (2015) have been important in drawing attention to the experience of parents whose children have been diagnosed with LKS. However, a more nuanced understanding of parents' experience of LKS is difficult to achieve from the small number of relevant studies. Consequently, it is pertinent to draw from other literature sources to hypothesise more elements of the parental experience which may be relevant.

Drawing together the medical and parental descriptions within the LKS research, particular features of the LKS clinical presentation are repeatedly highlighted. These features include; the rarity of the disorder; the regression or loss of previously acquired cognitive functions; altered developmental trajectories; and behaviour which can be experienced as challenging (Malvestio, 2010; Kuriakose, 2012). These clinical features have directed the continued literature review into more widely investigated disorders. The disorders selected for comparison do not represent an exhaustive list of all which share clinical features, but are rather a product of the comparisons made within the LKS literature and from discussions

with my academic supervisor who specialises in paediatric psychology.

### 1.7.1. Rarity

Cockerell et al. (2011) indicate many commonalities between their findings and research exploring other rare disorders. Although, by definition, rare disorders have a low prevalence, low prevalence does not result in a low impact. The total number of people affected by rare disorders within the EU is estimated to be between 27 and 36 million (Angelis, Tordrup, & Kanaet, 2015). Within the literature, importance is increasingly being placed on the individual experiences of daily life and acknowledging the physical, psychosocial, emotional and financial impacts of having a rare diagnosis (Zurynski, Frith, Leonard, & Elliott, 2008; Berglund, 2014; Pelentsov, Fielder, & Esterman, 2016 Dellve, Samuelsson, Tallborn, Fath, & Hallberg, 2006; Grut & Kvam, 2013; Wallenius, Möller & Berglund, 2009).

Firstly, the economic impacts related to the rarity of a disorder are associated with the challenges parents often have to make within their daily lives. Changing work schedules, unemployment, domestic responsibilities, healthcare costs and subsequent income changes have all been acknowledged as adding financial concerns for the family (Pelentsov et al., 2016; Zurynski, et al., 2008).

As seen within the studies of Cockerell et al. (2011) and Lemard-Reid (2014), parents of children with rare disorders often report a delay between initial symptoms and definitive diagnosis. Reasons why this gap exists include a lack of diagnosis, a lack of knowledge about rare diagnoses and their consequences, and a lack of communication between professionals (Berglund, 2014). Research exploring the subsequent impact of this 'diagnostic gap' on individuals and their families has presented a range of significant multidimensional challenges.

Primarily, the challenge in finding relevant medical information about the disorder makes it difficult to gain knowledge (Pelentsov et al., 2016). The implications of this, is that day-to-day management of needs can become challenging as treatment and caregiving advice may be unclear or unavailable (Berglund, 2014; Pelentsov et al., 2016).

Interactions with professionals have also been recognised to be unhelpfully affected by the rarity of a disorder. Grut and Kvam (2013) describe in detail the significant barrier this has on accessing adequate services as some services are reluctant to give support if the diagnoses are unknown to them. Furthermore, some professionals are reluctant to accept information offered to them by the service-user and consequently tend to base incorrect judgments and actions on their personal assumptions. In response to these findings, research, including the LKS study conducted by Cockerell et al. (2011), draws attention to the importance of professionals valuing the parents' experience. In some incidences parents may hold more knowledge than professionals and, if the professionals are uncomfortable with this insecurity and lack of knowledge, it might restrict helpful collaboration.

The physical challenges of accessing support and caregiving can then be further exacerbated by the emotional effects related to the rarity of the disorder. Research has reported a range of feelings that include uncertainty, stress, and humiliation within school and work settings (Berglund, 2014; Pelentsov et al., 2016; Dellve et al., 2006). Many parents also experience frustration, because of the perceived lack of knowledge of health professionals about the disorder (Berglund, 2014).

Parents of children with rare disorders often describe social isolation and strains on family relationships (Grut & Kvam, 2013; Pelentsov et al., 2016). Potential support gained through parent groups (if in existence) is often challenging to access as they can be geographically scattered (Pelentsov et al., 2016).

Within this context of rarity, the work of Dellve et al. (2006) highlights the features which promote parental coping. The key feature is the importance of increasing their perceived knowledge about the disorder. This is thought to shift the parent from using a passive to an active style of coping. Consequently, the parent is thought to develop and experience more helpful interactions with resources in the family, social network and wider society. Examples of this were, increased compliance with professional recommendations given and increased preparation

for meetings with healthcare professionals.

### 1.7.2. Regression of Abilities and Altered Developmental Trajectories

Observing the regression or loss of previously acquired cognitive functions of their child is understood to be a very distressing experience for parents.

Paediatric Acquired Brain Injury (ABI) is the most common cause of acquired disability in childhood, with an estimated 200,000 children affected in the UK (Antonini et al., 2014; Jordan & Linden, 2013). The impact is often that children present with significant cognitive changes, which consequently affect their ability to function independently, access education and engage in interpersonal relationships (Antonini et al., 2014).

As with LKS, children with ABI are also observed to have emotional, behavioural and personality changes (Malvestio, 2010; Antonini et al., 2014). Whether a result of the brain disturbance or the frustration associated with having profound difficulty in communicating, or both, the result is that parents often report that their children no longer behave as the child they once were.

Literature within this area has acknowledged that the regression in their child's ability affects the family, with some parents reporting significant distress (Antonini et al., 2014). Parents often take responsibility for tending to the physical and cognitive needs of their child and consequently experience caregiver and injury-related 'burden' (Brown, Whittingham, Sofronoff, & Boyd, 2013). Jordan and Linden (2013) explore in detail the concept of burden for caring a child with ABI. They describe it as multi-faceted, highlighting the significance of dealing with the unpredictability of their child's behaviour, watching their child frequently come across challenges, and experiencing grief as a result of 'losing' the life the family (including parent and child) could have had. Underpinning these experiences, Jordan and Linden (2013) also present the often contradictory emotions of love and frustration.

Another theme within ABI parental reports was the prolonged feeling of uncertainty (Kirk, Fallon, Fraser, Robinson, & Vassallo, 2015). As with LKS,

professionals are unable to give clear projections about the child's recovery trajectory and parents have to be aware that their child may or may not regain their damaged functions. Parents often have to acknowledge that the professionals themselves are working within a context of uncertainty (Malvestio, 2010; Kirk et al., 2015). Consequently, Kirk et al. (2015) identify the value parents place on professionals who are transparent about this restriction, as it provides a rationale to why information-giving was at times restricted.

The literature also addresses other themes such as: the difficulty of coping with the changes in their child; trouble adjusting to new family roles; coping in response to their child's changing needs over time; and a fear of future consequences of the ABI (Brown et al., 2013; Antonini et al., 2014; Jordan & Linden 2013). These experiences are understood to be further exacerbated by the challenges in accessing formal and informal support (Rashid et al., 2014; Jordan & Linden, 2013). In the context of frequent reports of negative reaction from others, including friends and family, Jordan and Linden (2013) suggest that it is not the brain injury that so adversely affects the mothers' emotional well-being, but the lack of individual and societal understanding.

A systematic review conducted by Rashid et al. (2014) explores the ways families cope with these challenges. Family cohesion was highlighted as a significant factor. Within increased cohesion leading to an increase in the parents perceived ability to cope and a reduced experience of stress. Furthermore, family cohesion was understood to be predicted by experiences of helpful social support.

In the work of Benn and McColl (2004), social support was also found to underpin more specific forms of coping. A perception-focused coping strategy was found to be implemented most frequently by parents of children with ABI, suggesting that the interpretation of the event was most significant. In the context of many ABI related stressors not being changeable, a positive appraisal or a meaningful interpretation of the child's presentation is thought to allow the family to view the challenges as more manageable.

### 1.7.3. Behaviour That Challenges

Frequent comparisons have been made within the LKS literature (Malvestio, 2010; Fandino et al., 2011) between Autism Spectrum Disorder (ASD) and LKS. Drawing attention to risk of misdiagnosis, researchers have outlined the commonalities of language regression and difficulties with social behaviour. Although a fundamental difference is that children with LKS preserve the desire to communicate and may be able to do so again once the active phase of LKS is over, the commonalities in presentation still provide an opportunity to explore and draw possible parallels with the more widely researched area of, parental experiences of ASD.

ASD is a developmental disorder involving abnormal communication, repetitive and restrictive interests, and impaired social functioning (Miranda, Tárraga, Fernández, Colomer, & Pastor, 2015). Well-documented research has highlighted the profound impact this can have on family life, explicitly outlining the experiences of the parents (DePape & Lindsay, 2015). Within this work, the multiple challenges and benefits of caregiving have been reported across the family life cycle (DePape & Lindsay, 2015; Ooi, Ong, Jacob, & Khan, 2016).

Caring for a child diagnosed with ASD has been shown to be linked to high levels of parental stress and depressive symptoms (DePape & Lindsay 2015; Kim, Ekas, & Hok, 2016). Within the complexity of symptoms, researchers identify a particular challenge being the behaviours which are disruptive and hard to manage (Ludlow, Skelly & Rohleder, 2012; Kim et al., 2016). Kim et al. (2016) specifically note a relationship within their research that presents increased severity of the child behaviour problems linked to mothers perceiving their child's daily life as less normal, lower confidence in managing their child's condition and family management requiring much more work. Within other research, parents also describe their frustration at not having time to themselves and other members of their family, as time is dominated by the demands of caring for their child with ASD (Ludlow et al., 2012).

In relation to the behaviour that challenges, parents report that one of the most difficult aspects is the social implications of their child's behaviour (Ludlow et al.,

2012). Whilst parents might have developed strategies to manage or accept behaviour that challenged, it was the lack of understanding and negative judgements from other that were particularly difficult. These negative critiques could be from strangers, friends or family and resulted in feeling like a failure or a 'bad' parent (DePape & Lindsay, 2015; Ludlow et al., 2012). Described by some as 'affiliate stigma' (Wong, Mak, & Liao, 2016), descriptions of isolation are common and even lead to parents reporting their discomfort in being out in public with their child as it is an invisible disorder, with people attributing parental blame (DePape & Lindsay 2015; Ludlow et al., 2012).

The impact on the spousal and sibling relationship has also been explored within the ASD literature. Presenting mixed findings, studies highlight that some spousal relationships experience challenges as a consequence of their child's presentation, while others found that the experience drew them closer together (DePape & Lindsay,2015). In the context of siblings, parents often reported spending more time with the child diagnosed with ASD and trying to compensate by reaching out to the other children when possible (DePape & Lindsay,2015).

As well as the challenges of caring for a child with ASD, research has addressed the ways in which parents cope. Ludlow et al. (2011) and McStay, Trembath and Cheryl Dissanayake (2015) draw particular importance to the role of social support, as it enabled the sharing of ideas and strategies to cope with the challenges. The work of Nicholas et al. (2015) also discusses the importance of assuming new roles, adapting future plans, and seeking support and solace through spirituality, to promote coping.

The wider meaning and positive identities associated with the parental role have also been discussed within the literature on coping (Nicholas et al.,2015; DePape & Lindsay,2015). Whilst acknowledging the struggle and revised expectations parents sometimes experience, Nicholas et al. (2015) discusses the less common topics of resilience and growth. Furthermore, the comparison with others can provoke an attitude of gratitude because their child's presentation could be worse (Ludlow et al., 2011).

## **1.8. Rational for Continued Research into the Parent's Experiences of LKS**

A more nuanced understanding of the parental experience of LKS has been developed through the incorporation of few studies explicitly focusing on parents' experience of LKS and the studies which have explored significant features that overlap with LKS. However, a number of reasons to continue exploring this area are apparent.

Firstly, although drawing from wider literature gives a valuable insight into potential commonalities of parental experience, there are limitations to the conclusions that can be made. Relying on research that is aimed at exploring the impact of other disorders can lead to unsatisfactory and simplistic assumptions about the specific parental experience of LKS. It is important to acknowledge that the parental experience of LKS may not simply be culmination of different experiences linked to separate features of the disorder, but rather a result of the unique interplay between the different clinical features. For example, might there be a difference in the parental experience of behaviour that challenges, when the disorder is relatively well-known and the professionals they interact with hold more knowledge about the expected development trajectory and treatment outcomes?

Secondly, when reviewing the LKS specific research, no studies have yet looked into the parents' experience within the United Kingdom. Consequently, it is unclear whether the findings to date are relevant to other social and cultural contexts. Furthermore, although these studies have used qualitative experience to allow for rich descriptions of parental experiences, all studies focused on a particular aspect of the experience.

Lemard-Reid's (2014) study focuses on the role parents can play in minimising the possibility of misdiagnosis, therefore the points emphasised and conclusions drawn within the study focus solely around the pathway to diagnosis. Although emphasising the importance of increased knowledge of this rare syndrome within the professional community, this study does not elaborate on the wider, post-

diagnosis parental experience. Furthermore, the study prioritises details of interaction with professionals, and features fewer descriptive accounts of parents' interactions with family or social networks.

Although Wairungu's (2015) case study looked beyond the process of diagnosis, this research was placed in the context of pedagogical challenges of supporting a child within an educational setting. Drawing attention to particular features of the LKS clinical presentation, the study states the importance of understanding the unique characteristics and challenges that could interfere with an equal education (Chapman, Stormont, & McCatherine, 1998). This specific focus on the education context and the case study methodology makes it challenging to draw generalisations beyond the individual child and family discussed.

Cockerell et al.'s (2011) research also explores experiences after the initial diagnosis, but focuses on the experiences of accessing the health and educational systems. Although gaining a rich wealth of information from the twenty participants interviewed, this study has again not facilitated an open exploration into the experience parents feel most pertinent for them. By limiting the range of context-specific issues that can be discussed, a more nuanced understanding of parents' experience of LKS is harder to achieve.

Therefore, further research into LKS allows parents to have continued opportunity to present the complexities of their experience of LKS and share a greater understanding of the disorder on a wider level that future parents and professionals may find beneficial.

### **1.9. Aims of The Present Study**

The current study aims to overcome the current gaps in our knowledge of parents' experience of LKS. Through the adoption of a qualitative design, it aims to enable parents to express a wide range of experiences associated with having a child diagnosed with LKS within the United Kingdom. This includes the challenges faced by parents and the factors that help them cope.

## **1.10. Research Questions**

The present study aims to address the following questions:

- How have parents experienced having a child with LKS throughout the course of the disorder?
- What are parents' past and current modes of coping?

## **2. CHAPTER TWO: METHODOLOGY**

The purpose of this study was to explore parents' experiences of having a child diagnosed with Landau Kleffner Syndrome (LKS). The relevant literature lacked descriptive accounts and focused predominantly on medical investigations. This chapter presents a rationale for the chosen research method, the process used to collect and analyse the data, the procedures for recruiting participants and the ethical considerations.

### **2.1. Qualitative Research**

The majority of research exploring LKS has utilised a quantitative, objective approach. Focusing on quantifiable data, studies have uncovered causal relationships which give valuable insight into this rare syndrome and its devastating consequences. Most significantly they have identified the relationship between seizures and language dysfunction (Tuchman, 2009), the syndromic definition (Miguel et al., 2011) and the prognostic indicators (Arts et al., 2009).

Whilst acknowledging that quantitative study has been able to provide insights into the understanding of LKS, it is less able to explore the neglected area of human experience. A qualitative approach provides the opportunity to gain unique insights and understandings (Willig, 2008). Furthermore, drawing knowledge from individuals who have experienced the phenomena allows context to be acknowledged and valued.

In the absence of extensive research into the parents' experience of having a child with LKS, a qualitative approach enables a rich description of the particular challenges faced and the resources needed to cope.

## **2.2. Epistemological Position**

Carter and Little (2007) highlight the importance of establishing an epistemological position when conducting qualitative research. Defined as “the study of nature of knowledge and justification” (Schwandt, 2001 p.71), it allows the researcher to locate their source of knowledge and consider the extent of its reliability (Harper, 2012; Willig, 2008). In turn the epistemological position shapes the choice of methodology and method employed to answer the research questions.

To address the research questions of the current study, a critical realist epistemological stance was taken. Positioned between a realist and relativist epistemological perspective, it recognises that our observations and knowledge about the world are not objective and are instead influenced by the participant and researcher’s perceptions (Willig, 2008). Taking a critical perspective on taken-for-granted knowledge, our perception of the world can be understood as developing out of multiple contexts, including the societal and historical setting.

In asking parents about their experience of being a parent to a child with LKS, I am assuming that their experience is not an uncovering of the ‘truth’ which can be quantified, but rather an expression of an experience which is contextually and historically specific (Harper & Thompson, 2011).

## **2.3. Method**

### **2.3.1. Choosing A Qualitative Method**

The method chosen to answer the research questions is shaped both by my epistemological and professional position. An assumption that people experience a ‘reality’ which is influenced by significant factors that they may not be entirely aware of, is a key overlapping theme of the critical realist position and theories within clinical psychology.

There are many qualitative methods available for the exploration of the ways in which people make sense of world(s) they inhabit (Harper et al., 2011). The suitability of a particular method of analysis is dependent on the nature of the exploratory research task. The approaches considered in the current study include interpretive phenomenological analysis, narrative analysis, grounded theory and thematic analysis.

The overarching feature of these approaches is their ability to present individuals' understanding of the world they interact with and the experiences they have had. These approaches are described in turn below, followed by a rationale as to why Thematic Analysis was felt to most appropriately address the study's research questions.

#### *2.3.1.1. Interpretive Phenomenological Analysis*

Interpretive phenomenological analysis is an approach which explores the meaning people make of their experiences and how they relate to the world (Smith, 1996). Exploring both the social and personal world of the person, verbatim transcripts of individual interviews are analysed in detail. The aim is not to collate facts about what happened, but rather to explore the meaning and significance of an event for an individual (Larkin & Tompson, 2012). Consequently, the findings from these methods are thought not to be generalisable.

#### *2.3.1.2. Narrative Analysis*

Narrative analysis is also interested in the ways people interpret the world around them, but places emphasis on the stories people tell themselves and others, and how they told or communicated (Murray & Sargeant, 2012). As with interpretive phenomenological analysis, this approach does not view the participants as representative of a wider population. Results are a unique account of how someone constructs their experiences and cannot be generalised.

#### *2.3.1.3. Grounded Theory*

Although there are different versions of grounded theory (Glaser & Strauss, 1967), its overarching feature is the ability is to develop theory out of empirical

data. Utilising broad and open-ended questions, the patterns and meanings within people's experiences are explored (Tweed & Charmaz, 2012). Focusing on social or psychological processes, researchers categorise the data with the aim of identifying underexplored links and relationships (Willig, 2008).

#### *2.3.1.4. Thematic Analysis*

Thematic analysis is a method designed to identify and analyse patterns of meaning within complex or sensitive data (Braun & Clarke, 2006). The aim is to uncover salient themes which represent the nature of a specific group's experience or understanding of a phenomenon (Joffe, 2012). Both the meaning participants give to these themes and the influence of broader contextual factors are considered, giving rise to the identification of manifest (directly identifiable) and latent (implicit) content (Barker, Pistrang & Elliot, 2002).

Although the current study is an exploratory study, the emphasis is to highlight the experiences encountered by parents. A detailed account of an individual's reflections on their experiences or how they communicate their experiences is not prioritised. Additionally, the current study aims to analyse data from across a sample, leading to a broader focus. For these reasons a narrative analysis and interpretive phenomenological analysis are not appropriate for this study.

The current study also aims to gain an understanding of the parents' experiences without the expectation of linking themes or of developing theoretically driven relationships. For this reason, the implementation of a grounded theory approach was also thought to be unsuited to the current study and its research aims. In conclusion, the greater flexibility of a thematic analysis was felt to be the best fit to accommodate the questions being proposed by the current research.

#### 2.3.2. Rigour, Validity and Reflexivity

Whilst thematic analysis does not explicitly prompt the researcher to consider their role as an active participant within the process of research, Braun and Clarke (2006) highlight the importance of doing so. Engaging in reflexivity is crucial in conducting rigorous and valid qualitative research (Harper, 2012) and allows the researcher to acknowledge that there is no single objective truth or

reality to be discovered (White, 1992). Instead, the researcher draws attention to their own contribution to the construction of meaning and how this may influence the research itself. Themes drawn from the data are understood to be influenced by the researcher's existing knowledge, prior research interests, values and experience of the research topic (Ely, Vinz, Downing & Anzul, 1997).

Yardley (2000) suggests four principles be adopted to ensure validity within health psychology research: 1) sensitivity to context; 2) commitment and rigour; 3) transparency and coherence; and 4) impact and importance. The ways in which the current study endeavours to meet these criteria of rigour, validity and reflexivity will be outlined within the discussion chapter.

### 2.3.3. Method of Data Collection

As the aim of the current study is to explore parents' experience of having a child with LKS, the method of data collection needed to encourage the participants to express themselves openly. There are a number of approaches which enable this, whilst being consistent with a qualitative methodology and critical realist epistemological position. The current study considers the appropriateness of questionnaires, focus groups and verbal interviews, before concluding that verbal interviews are the most appropriate method for the current study.

#### 2.3.3.1. *Questionnaires*

Self-administered qualitative questionnaires are able to obtain large amounts of information from a broad geographical area in a relatively short amount of time. This is a useful feature of the approach, when considering the fact that rarity of LKS occurrence results in affected families being geographically dispersed. With the use of open-ended questions, questionnaires can also facilitate participants expressing their views in response to the question asked without any influence or clues from the interviewer (Robson, 2002). Furthermore, they encourage honesty from participants as anonymity can be ensured (Robson, 2002). This is a valuable factor when the participants are being asked about potentially emotive or challenging subjects such as the feelings towards their child's symptoms of LKS or their interactions with professionals.

However, there are limitations to the use of self-administered questionnaires. The lack of supervision means potential misunderstandings of questions may go unnoticed. To avoid this, the researcher may try to simplify questions, potentially limiting the depth of responses gained (Robson, 2002). Questionnaires are also known to have a relatively small respondent rate (Robson, 2002). In the context of LKS being so rare, the population being accessed is already relatively small, therefore the responses are not only at risk of being simplistic in content but also small in numbers.

#### *2.3.3.2. Focus Groups*

Focus groups produce data through creating opportunity where interactions between the group members occur. Providing a space where people listen and reflect, individual responses are thought to become refined and more considered (Finch & Lewis, 2002, p.170-191). Within this structure, the researcher becomes less influential as a more naturalistic setting is developed between the participants (Finch & Lewis, 2002).

An obstacle to utilising this method is the potential reluctance to share minority views and experiences within a group (Frith, 2000; Kreuger, 2008). Another challenge is the logistical requirements needed to facilitate the meeting. In the context of LKS, the rarity of the disorder means the population is geographically dispersed and may limit potential attendees.

#### *2.3.3.3. Verbal Interviews*

The key element of interviews is their focus on the individual. They allow complex ideas to be discussed and the interviewees' experiences to be located within their personal context. The three most commonly used interview formats are structured, semi-structured and unstructured. Defined by their reliance on predetermined questions, each one gives a different level of freedom to the interviewer (Robson, 2002).

Semi-structured interviews aim to be a mid-point between structured and unstructured formats. Although the interviewer has a guideline of topics and questions they would like to cover, they have the flexibility to ask additional

questions based on unexpected or unanticipated answers. This method ensures depth to the data because the researcher has the opportunity to clarify and understand the significant detail of the experience being shared (Arthur & Nazroo, 2002). Individual interviews can also take place in locations where the participant feels most comfortable, promoting open dialogue.

Benefits of this method are restricted when considering the time and resources needed to conduct them (Kvale, 1996). The researcher is also more influential in this setting and potentially risks creating response bias or privileging their own agenda (Arthur & Nazroo, 2002).

Despite these challenges, the ability of semi-structured interviews to generate rich detailed accounts of experience for a small, broadly distributed population, made this the most appropriate method to use within the current study.

## **2.4. Selection and Recruitment of Participants**

### **2.4.1. Sample Size**

Qualitative research typically requires fewer participants than quantitative research, as the priority is to gain a rich understanding of a subjective experience. The current study aims to have a sample size of six to twelve participants, based on literature exploring the amount of data needed to reach 'data saturation' (Burmeister & Aitken, 2012) and uphold the study's desired strength of claim (Charmaz, 2006 p. 114).

Data saturation refers to when further sampling is not deemed likely to yield significantly more novel information and substantial repetition is seen within the data. However, drawing from the work of Mason (2010), Fusch and Ness (2015) present the complexity of defining data saturation. They highlight the subjective nature of this desired goal within qualitative research, acknowledging the influence of the methodology chosen and the researcher's 'personal lens'. Fusch and Ness (2015, p.1410) suggest that the better a researcher is at reflecting on

their own frame of reference, the better they are able to record, interpret and reflect on the experience of others.

Whilst recognising this complexity, researchers have agreed on some general principles to indicate when data saturation is reached. These include: when further coding is no longer feasible; when no new themes are emerging (Guest et al., 2006); and when there is enough information to replicate the study (O'Reilly & Parker, 2012; Walker, 2012).

Guidance on sample sizes has emphasised the importance of the researcher's aims, drawing attention to the study's desired strength of claim (Charmaz, 2006 p.114). The current study can be considered to hold modest claims and can therefore be understood to reach data saturation more quickly. Baker and Edwards (2012) suggest a sample size of six to twelve when the population accessed is under-assessed and valuable. This is relevant in the context of minimal qualitative research having been conducted about the rare disorder, LKS.

#### 2.4.2. Participant Identification

The study aimed to identify participants who had a wide range of experiences related to parenting a child with LKS. Participants were initially identified from a database of current and ex paediatric patients who had been diagnosed and/or treated for LKS at a specialist Paediatric London Hospital. This database included the contact details of the child's parents or carers. Through the exploration of relevant literature into parents' experiences of other childhood disorders and clinical discussions with my field supervisor, who has frequently worked with families affected by LKS, an inclusion and exclusion criteria was developed.

The inclusion criteria selected parents whose children were no longer in the active phase of LKS. This means that their child was no longer taking steroid medication for LKS, no longer showing Electrical Status Epilepticus in sleep, and whose language abilities had stabilised.

The exclusion criteria lead to parents not being approached if their child had recently gained a diagnosis of LKS and/or whose children were considered to still be in the active phase of the disorder. This active phase included showing sleep activation discharges on a sleep electroencephalogram (EEG), taking steroid medication for LKS, and presenting with fluctuant language ability.

The decision to only invite parents whose child had a historical diagnosis and therefore was no longer in the active phase, arose through an assumption that these parents may have experienced many aspects of parenting a child with LKS, including post recovery. Parents whose child had recently gained a diagnosis may be preoccupied with negative experiences (e.g. shock and devastation) (Sen & Yurtsever, 2007). This potential bias within the data was thought to limit the opportunity of highlighting other aspects of a parent's experiences (e.g. resources drawn on for support or improvement in their child's prognosis).

Whilst outlining the rationale for the criteria developed, the study also acknowledges the potential limitations to adopting these criteria. For example, parents whose children received a diagnosis historically may not be able to accurately recall the subtle detail of their experiences. Furthermore, their interactions with health and educational services may have changed over the years, perhaps limiting the generalisability to parents who are recently affected.

Overall, however, it was felt that the advantages of the parent being able to draw from a breadth of parental experiences, would outweigh the potential limitations outlined.

#### 2.4.3. Recruitment Process

Recruitment was conducted over a three-month period from November 2016 to January 2017. Out of ninety three children and their families listed within the database, twenty nine were identified as meeting the inclusion criteria. Fifteen families of those children who met the inclusion criteria were selected by random and contacted by invitation (see appendix B). This invitation letter outlined the research project and what their participation would require. If a parent was willing to participate, they were asked to opt in, via email, letter or telephone to myself.

Participants who did not make contact by the opt-in date (two weeks from the date the letter was sent) were contacted by telephone to clarify that they received the letter. The telephone call also enabled the parents to discuss any possible questions or concerns they may have about participating. As sufficient participants were not identified, a further five families were contacted in the same way to enable the threshold of participants needed to be reached.

#### 2.4.4. Participant Details

Drawing from the guidelines provided by Sanders, Kitzinger and Kitzinger (2015), basic demographic details of the participants and brief stories about each family were collected and are provided below. It is hoped that this anonymised information provides the reader with context that facilitates a richer understanding of the data. Due to the rarity of LKS, in-depth participant descriptions will not be available because it may make participants identifiable.

##### 2.4.4.1. *Participant 1*

Participant one was a forty-five-year-old woman, whose child with LKS was nineteen at the time of the interview. She described herself as a housewife and reported GCSE's to be her highest level of education. She lived with her partner, who was the father of all three of their children. Their son with LKS was the oldest of his siblings. At the time of the interview, the participant reported her son to have residual language difficulties, severe memory difficulties and to be very shy.

##### 2.4.4.2. *Participant 2*

Participant two was male in his late forties, whose daughter with LKS was eighteen at the time of the interview. He was married to participant three, and together they had three children. Their daughter with LKS was the youngest of their children. Participant two described himself to be self employed and reported his highest level of education to be GCE's. He described his daughters difficulties at the time of the interview as; very vulnerable, defiant, difficulty maintaining boundaries and difficulty with word finding.

#### *2.4.4.3. Participant 3*

Participant three was a 49 year old female, who was married to Participant two. She described her eighteen year old daughter with LKS, as having the same difficulties at the time of her interview, as her husband. She described herself as self employed, primarily fostering children with learning difficulties.

#### *2.4.4.4. Participant 4*

Participant four was a 49 year old woman whose son with LKS was twenty years old at the time of the interview. She worked part time and reported her highest level of education to be the equivalent of A levels. She was married and had two children who she lived with. Her son was the eldest child. At the time of the interview, she reported him to have no cognitive or emotional difficulties.

#### *2.4.4.5. Participant 5*

Participant five was a woman in her sixties, who had four children. Her son with LKS was the youngest and was 23 years old at the time of the interview. All children had moved out of the family home, and she and her husband were now retired and lived alone. She had previously worked as a medical professional and archived a Masters degree. Her son with LKS was reported to have speech and language difficulties and a slower than average processing speed.

#### *2.4.4.6. Participant 6*

Participant six was the husband of participant five. He was also a retired medical professional, with the highest level of education being at a doctorate level. He reported his son to have the same difficulties as described by participant five.

#### *2.4.4.7. Participant 7*

Participant seven was a 43 year old female, who was employed full time. She was married and lived with three of her four children. Her duties included helping other families whose children had learning difficulties. Her daughter with LKS was twenty one years old and was the second eldest. At the time of the interview she

was described as having difficulties with learning and reported to need day to day support with independence.

#### *2.4.4.8. Participant 8*

Participant eight was a female in her mid Fifties, whose son with LKS was sixteen at the time of the interview. She described herself to work part-time, where she supported children who had learning difficulties. Her highest level of education was a masters. She was married and lived with her two sons. Her son with LKS was the youngest. She described her sons difficulties at the time of the interview as; having slight residual language difficulties, slower processing speed and minor word finding difficulties

## **2.5. Ethical Considerations**

It is imperative that researchers are guided by ethical guidelines throughout the different stages of the study (Thompson & Chambers, 2012). A high standard of professionalism is expected from psychologists across all their roles, including that of a researcher. The British Psychological Society's (BPS) ethical guidelines were used to consider how the current study could potentially create vulnerable situations where harm could be caused to participants and the researcher, and how this would be managed (BPS, 2010).

### **2.5.1. Potential Distress and Safeguarding**

In the context of this study it was crucial to consider how I might work sensitively and safely with parents who had experienced challenging and upsetting times. Although the interview agenda did not intend to cause distress, it was expected that the information shared might include personal and emotive subjects. I approached and responded to these topics respectfully and thoughtfully, drawing from my skills as a Trainee Clinical Psychologist to assess whether further support would be needed (Thompson & Chambers, 2012). Opportunities to ask questions or have breaks within interviews were offered to all participants.

Protection for the researcher was also considered, with my supervisor having

knowledge about when and where my interviews were taking place. Regular supervision was also arranged throughout the data collection period. This space provided the reassurance that any vicarious distress I might experience or safeguarding concerns I might encounter, would be supported. Supervision additionally gave rise to reflections about my interactions with participants. Themes of power, professional roles held and challenges to interviewing were discussed, in order to continually develop and improve my skills as a researcher.

#### 2.5.2. Ethical Approval

Ethical approval for this study was initially sought and obtained from Great Ormond Street Hospital's Clinical Research Adoptions Committee on the 23rd of May, 2016 (appendix C). Ethical approval was then sought and obtained from the Westminster Research Ethics Committee on the 30<sup>th</sup> of September, 2016 (appendix D). Subsequently, ethical approval was sought and obtained from the Health Research Authority on the 28<sup>th</sup> of October, 2016 (appendix E).

#### 2.5.3. Informed Consent

Before any interviews were conducted, all participants were given an information sheet describing the study and clearly outlining what their participation would involve. This included an assurance of anonymity and confidentiality (see appendix F). Participants were also made aware of their right to withdraw from the study and were given the opportunity to ask any questions or share any queries they had about the study. All participants were requested to sign a consent form before they participated (see appendix G).

At the end of the interview, participants were given a debrief sheet, thanking them for their participation and giving details of who to contact if they had any questions or concerns (see appendix H).

#### 2.5.4. Confidentiality

Anonymity and confidentiality was assured, with all interview recordings password protected and all transcripts anonymised with the use of pseudonyms. Demographic information requested about the participant and their child was also anonymised. Boundaries of confidentiality were outlined before the interview took

place (e.g. discussion with my supervisor if there are concerns of risk to themselves or another.) Interview transcripts were only available to the researcher, supervisor and examiner. They were stored in a locked cabinet, with access only available to clinicians involved in the research. All interview recordings will be deleted after 5 years, as outlined by the Data Protection Act (1998).

## **2.6. Interview Process**

### **2.6.1. Materials and Interview Schedule**

An Olympus digital voice recorder (WS-450S) was used to record the interviews. In full view of the participants the recorder was placed between myself and the interviewee. A semi-structured interview was conducted, using an interview schedule (see appendix I). The initial questions informing the interview schedule were developed out of a literature review and the aims of the current research project.

### **2.6.2. Pilot Interview**

The questions within the interview schedule were initially posed to one participant who met the inclusion criteria. This was to assess whether revisions should be made to my interviewing style or questions, to ensure the participants responses met the study's objectives.

The pilot interview confirmed that I did not need to make changes to my interview schedule, as the questions prompted the participant to give a detailed and rich account of their experiences. This first interview also reduced my initial concerns that I may find it challenging to make the necessary shift away from my clinical role as a Trainee Clinical Psychologist, to positioning myself as a researcher. As the stories were predominantly historical, I found it comfortable to prioritise listening and facilitate open conversation and not to provide support.

### 2.6.3. Research Interviews

The participant always chose the location, with my only request being that it should take place in a quiet environment. Eight participants were interviewed, all of whom requested that the interview be conducted within their homes. It seemed this familiar setting encouraged the parents to feel at ease.

I started each interview with an introduction explaining who I was and a recap of the purpose of the interview. I assured confidentiality and reminded them that the interview would be recorded. After the participant had agreed to take part in the study and signed the consent form, the semi-structured interview began.

I adhered to particular interviewer behaviour such as posing questions in a non-judgmental way, actively listening, having neutral body language and presenting as engaged and interested in their conversation. The interviews lasted between forty five minutes to 1 hour and 15 minutes and were concluded by thanking them for their participation and giving them a copy of the debrief sheet.

## **2.7. Process of Analysis**

Once all interviews were completed, the recorded interviews were transcribed verbatim using Parker's (2005) transcription convention (see appendix J). Both an inductive (data-driven) and deductive (theory-led) thematic approach was used (Braun & Clark, 2006). Incorporating these approaches enabled me to utilise my knowledge from pre-existing theories to develop my sensitivity to subtle features of the data, whilst giving space for unexpected themes to emerge (Joffe, 2012). Drawing from Braun and Clark's (2006) six-phase analytic process, thematic analysis was conducted with the data. These phases were not implemented chronologically, but moved through flexibly, allowing some phases to be repeated when necessary.

### 2.7.1. Phase One: Familiarisation with the Data

Familiarisation with the data began at the early stage of data transcription. By listening to the interview recordings and typing them up onto a Microsoft word

document, I was able to actively engage with the data (Braun & Clark, 2006). My initial thoughts, and recognition of patterns and ideas, were subsequently noted whilst reading and re-reading the transcript (see appendix K for an excerpt of a transcript).

#### 2.7.2. Phase Two: Generating Initial Codes

Working systematically through the transcripts, initial codes were developed from the notes I made whilst becoming familiar with the data. These codes represented subjectively meaningful groups into which the data could be organised. After each transcript had been initially coded, excerpts were collated. This process allowed the data to be compared. It also assured that the codes were consistently assigned. A list of codes was then generated (see appendix L).

#### 2.7.3. Phase Three: Searching for Themes

The final codes were repeatedly read and explored, to identify how they may be combined to develop overarching themes and sub-themes. This phase ceased only once data saturation was thought to have occurred, with no new codes or themes emerging (Guest et al., 2006) and there was enough information to replicate the study (Braun & Clark, 2006).

#### 2.7.4. Phase four: Reviewing Themes

During this phase, the themes were initially reviewed by re-reading the coded extracts within each theme. If inconsistent patterns were observed across the grouped codes, adjustments were made. During this phase, codes could be moved to more appropriately matched themes or sub-themes, or new themes could arise (see appendices M-N).

After this stage, themes across the entire data set were then reviewed. The aim was to ensure that the themes represented the entirety of the data. All the transcripts were re-read and themes were further adjusted.

#### 2.7.5. Phases Five: Defining and Naming Themes

The final themes and sub-themes were revisited, allowing their relationship and relevance to my research questions to be established. This phase also continued

to ensure that all significant elements within the data had been captured within the clearly defined themes.

#### 2.7.6. Producing the Report

Within the Results chapter of this thesis the detailed thematic description of the data will be presented. Anonymised with pseudonyms, quotations from the interviews will be used to exemplify the context and meaning of themes and sub-themes.

### 3. CHAPTER THREE: RESULTS

This chapter will examine the themes and sub-themes derived from the data analysis. Two themes were constructed and are presented in Table 1. Extracts of data will be used to support each theme with the descriptive terms 'all', 'most, and 'some' used to indicate level of participant response. Small adjustments have been made to the text to improve readability of the quotes (Parker, 2005). This includes using (...) to indicate where words have been omitted and square brackets for the inclusions of text for explanation purposes e.g. [text]. To protect the anonymity of participants, participant numbers have been given to each parent and their child's name replaced with a X. Further analysis of the data in relation to existing literature is discussed in the following chapter.

Table 1. Themes and Sub-themes

<b>Theme</b>	<b>Sub-theme</b>
1. Challenges to Coping	a. Witnessing The Change: Loss Of The Child
	b. Rarity: No One's Listening
	c. Implications For The Family: The Knock On Effect
2. Evolution Of Family Roles And Ways Of Coping Over Time	a. Advocacy: They're Not Gods
	b. Finding Strength and Power in Personal Resources
	c. Acknowledging the positive
	d. Making Meaning"
	e. One day at a time

#### 3.1. Theme 1: Challenges to Coping

This was one of the most frequently mentioned themes, addressed by participants in nearly every interview. It encompasses the challenging experiences that were related to specific features of the LKS presentation and the

implications this had on the parent and family. This theme is comprised of three sub-themes: Witnessing the change: loss of the child, Rarity: No Ones Listening and Implications for the family: The Knock on Effect.

### 3.1.1. Sub-theme 1a: Witnessing the change: loss of the child

LKS is characterised by a deterioration of language ability after a period of typical development (Caraballo et al., 2014). The varied LKS presentation also leads some children to present with overt seizures and extreme changes to behaviour (Caraballo et al., 2014). Mentioned by most participants, this dominant sub-theme captures the experience of witnessing this deterioration occur. For some parents, loss of language skills was the first sign that their child was unwell.

*“When he was about three and a half is when I first started noticing difficulties. Particularly in his speech. It was sort of erratic and garbled sort of nonsensical words...he left nursery speaking full complex sentences, understanding things. And in that one summer he just started losing masses of language.” (P8: 9-13)*

Most parents described a significant change in their child’s behaviour, providing a vivid and emotive picture through stories of their child behaving like they never had before. The stories also exemplified the extreme nature of the change with frequent mentions of the child acting aggressively or engaging in risky and dangerous acts.

*“He was hyperactive, he was impulsive and saw no consequence... He tried to stab his dad.... I could have a pot on the stove with water, whereas before he would have never thought of going to the stove ... he was drawn to anything with danger.” (P1: 375-379)*

The changes observed in the child were not just in relation to the initial onset. In the context of treatment outcomes or the general development of LKS, most parents also highlighted the difficulty of watching their child acquire and lose skills across the course of the illness.

*“So you're hoping for that [recovery] to happen and then it doesn't. And then there is something else. Hoping a regression doesn't happen, then it does. And it really did feel like snakes and ladders” (P8: 429-432)*

*“We got a tutor for her in the summer holidays to keep on top of this [deterioration of skills]. She'd done it all and then the next thing she started back at school and then it had all gone. All this work that we'd paid in, it had all gone.” (P7: 40-43)*

In the context of witnessing these changes, some parents made parallels with losing their child. The loss of the child was mentioned in the context of unfamiliar behaviour, presenting them with a child that they did not recognise.

*“I had this little petite blonde with pure straight hair. To this different child, with ginger curly hair...I had to grieve my little girl. The girl I had and expected. A gorgeous little girl.” (P7: 75-79)*

The emotional impact of watching their child change and become unwell led most parents to experience anxiety, fear and distress at some point during the course of the illness. For some parents these experiences were connected to not knowing the cause of their child's presentation and therefore caused initial concerns that there may be a serious underlying reason that may lead their child to die.

*“And I have a great GP so she asked what was my biggest fear. My biggest fear was he actually had a brain tumour or something because it was clearly getting worse.” (P8: 23-24)*

*“...I'd be calling the ambulance in the night because he couldn't breathe. He'd go blue. It was every single night. We'd have to take it in turns to see him because we were worried he might die. It was that bad.” (P4: 72-73)*

For some participants, the emotional experience also included confusion. As some of the children did not experience observable seizures, the child's presentation was particularly difficult to understand.

*“And it was such a difficult illness to get your head round because he never had dropped down seizures. Never had seizures that other people would see or I would see. So you literally had no idea.” (P8:292-294)*

As well as experiencing distress for themselves, some participants explicitly shared concern for their children's emotional wellbeing. This concern was most typically in relation to the social impact of changes to their cognitive abilities and physical presentation.

*“... I always used to go up there [school] at lunchtime. She was always sitting at this bus stop and no one was playing with her. But at that time she was on steroids and she's really fat... So she's gone from being a real skinny little thing to this really big thing that she could hardly walk. So it was probably scary for the other children as well because all of a sudden X had completely changed and they probably didn't recognise her.” (P2: 133-138)*

### 3.1.2. Sub-theme 1b: Rarity: No One's Listening

LKS is recognised as a rare syndrome (NIH, 2015), with the incidence calculated to be approximately one in a million (Kaga et al., 2014). This sub-theme explored the parents' experiences relating to the rarity of the LKS presentation. Expressed through the stories, the codes for this sub-theme related to the potential barriers and challenges many parents faced as a consequence of themselves, their social networks or professionals having little knowledge of the disorder.

As a consequence of the unfamiliar presentation of the child's symptoms, some parents mentioned the broad range of explanations initially given or investigated. These included both emotional and biological.

*“So we start a food diary. Literally every part of his life. The washing powder I used. What we done that day. What he ate. Everything. Everything was written down because they said it wasn't a seizure.”*  
(P1:53-55)

The journey to gaining a diagnosis was negatively affected for most participants by the rarity of LKS. A few parents described initial medical investigations as inconclusive, while others shared their experience of their child initially receiving an inaccurate diagnosis.

*“...they did some tests and they said he might be autistic. I said, he's not autistic. I said because he was fine. And now he's not fine. and if you're autistic I think you're born with it. It's something you would know from a very young age.”* (P4: 30-33)

Some parents were first told their child had epilepsy before the establishment of a diagnosis of LKS. This caused contrasting parental reactions from initial feelings of reassurance to emotions of upset and devastation.

*“I went in. He was this big Doctor. Then he said, she's got epilepsy, don't ever leave her alone, never let her ride a bike and never let her swim. And sent me out the door. I wasn't that old, I mean I was in my early twenties so I was. And then from that, there was floods of tears.”*  
(P7: 48-51)

In relation to the their perceived lack of knowledge, a few parents highlighted the shock of hearing their child had a form of epilepsy.

*“We didn't put it down to seizures whatsoever because as a family or as people that knew a little bit about seizures, we thought they were going to present themselves in a completely different way.”* (P1: 14-16)

In response to parents gaining a final diagnosis of LKS, most parents reacted with concerns relating to the rarity of the disorder. Primarily, parents felt confused about what the diagnosis meant and found it unhelpful to have so little information available. The diagnosis also seemed not to curb parent's worry about the future, as the recovery trajectory could not be stated.

*"Well I'd never heard of it. The Internet was all relatively new. No literature in the hospital. Didn't know repercussions, you knew only what the doctors were telling me" (P1: 262-264)*

*"We still dreaded his future. I mean I never thought we'd get our lives back together. Ever." (P1:331)*

For one mother, the LKS diagnosis was experienced positively as it relieved concerns about a feared brain tumour. However, this did not prevent her from experiencing the same worries about the future as the other participants had shared.

*"So when they did actually say it was LKS it was a real relief to get a diagnosis. It wasn't a brain tumour... So it was a relief to get the diagnosis. It's also really really scary" (P8:63-66)*

*"I did have hopes for the future. But there were hopes. I had no guarantees, had no sense that everything would be fine. Even when he was getting a lot better I still didn't quite trust it." (P8:353-354)*

The variety of presentations seen with children who have LKS, made it hard for some parents to see commonalities between their experiences and others. This was seen as a barrier to finding interactions with other parents within the LKS community helpful. Parents noted a number of influential factors that made relating to each other challenging; the stage of their child's illness (e.g. beginning or post recovery); the child's presentation (e.g. presence of observable seizures or behavioural challenges); the family dynamic (e.g. single parent or number of

children); and logistical challenges (e.g. living in a rural area versus near the specialist hospital).

*I got a note home one day saying, I thought you would like to know but we've got a new student at school who has the same condition and we thought maybe it would be nice for you to meet up. And I was like oh wow and I was so excited. And when I met her they was so different and he never had a seizure in his life. So I just sat there and I was listening to this woman who thought her whole world to coming to an end. And I'm looking at her thinking you've got no idea." (P2: 403-408)*

Within the stories shared, parents highlighted their interactions with friends and people within the general community. Negative experiences were mentioned slightly more frequently and touched on topics of exclusion and isolation. In the context of others' fear and social judgment, parents talked about both the withdrawal experienced from others and the withdrawal of themselves from others. A few parents also addressed the invisibility of the LKS and suggested this may have made it harder for other people to understand their child's presentation and led to people to take a more judgmental position.

*"I think what's interesting about having anyone in the family that's not a hundred percent is if you can't see it you can pretend it's not there, but when you can see there's something wrong, whether it's behavioural or whether it's physical with the bloating and not looking quite right, I think it becomes a much harder thing to bear because you're witnessing other people's reactions to that. And I think that can become quite a burden..." (P5: 391-395)*

*"I mean for a whole year I stopped taking her out because when I did go out, all the people stared. I always remember when I did, this woman turned around and said, oh you've got a naughty little girl there. I went, no, I've got a poorly little girl." (P7: 140-142)*

For other parents, the wider family, including the child's grandparents, were seen as unhelpful. Their reasons varied and included; living far away; making judgements; and being fearful and having misunderstandings about their child's presentations. These features of their experiences made receiving support from some people within their family, unhelpful.

*"I had no one to look after X because my mum and dad, they refused. Because they were scared. They didn't refuse because they didn't love him. Because my mom was scared that he's going to die on her. So she said she can't take responsibility." (P1: 438-441)*

Parents frequently described their experiences of interacting with professionals associated with health and social care services. Negative experiences with professionals included experiencing judgement, their concerns being dismissed and not listened to, lack of professional knowledge about LKS, inconsistent staff and unhelpful strategies provided.

*"I found it frustrating that people come in with all these thoughts. They don't know your son. You're telling them and no one's listening. I found people coming to the home intrusive, I felt like a failure when the behavioural therapist's tactics weren't working, but I knew what he was doing wasn't his fault. Something needed to be fixed." (P1: 418-422)*

*"We had someone come out from social services to try and do things with him. But it was just a complete waste of time. X had got bruises and they were trying to accuse me of hurting him. Well this was a child, I'm not joking you, if I was in the back of X he would fall forward, if I was the front of X he would fall backward. I couldn't be everywhere, he was a big boy...I told them in no uncertain terms that I never wanted them back again. Because I'm trying to fight for him to get better, I'm not trying to hurt him. It's strange how people can be...people like that I just kept them out of the way. Keep focused." (P4: 506-514)*

*“And I'm only a mum, I don't have all of these degrees. I just knew that I was living with this nightmare of a child. And the Professor saying to me, come back in six months' time, you try doing it for a day. You can't look at this child in this room. There was no connection. It was like he was talking to me about a child that wasn't even in the room. To me he wasn't observant enough.” (P4: 455-459)*

The severity of LKS meant that the children of all participants interviewed had engaged with medical treatment. The child of one participant had undergone brain surgery. Most parents expressed negative emotions related to their child's receipt of treatment although a few related positive experiences following a successful outcome. The negative experiences shared related mostly to the rarity of LKS and the lack of a clear treatment pathway.

*“...try this drug and come back in six months...I don't need that. I just knew it. I knew the drugs weren't going to work. I don't want him to have epilepsy, I want the epilepsy to stop. I don't want to keep giving him drugs. I didn't want him to even start taking the drugs and the drugs weren't working. So why give him more. I just thought, no I'm not doing it. I'm just not doing any more. And just to see him as zombie.” (P4: 372-376)*

*“She was a guinea pig as far as I was concerned. Even with the professor, just give her some of these, just give her more tablets.” (P7: 99-100)*

The rarity of the LKS presentation and the lack of knowledge professionals held, led to mixed responses when accessing educational support for their child. Some parents commenting on both the 'fight' to access appropriate support and the great benefits seen when the support had been perceived to be helpful. The challenges experienced while interacting with educational settings included: selecting the most appropriate school for the child's academic and behavioural needs; gaining a Statement of Special Educational Needs; recommendations not

being followed up; and finally, professionals lacking knowledge and skills to support specific needs of their child.

*“So the only thing I'd say is once you fought the medical battle it's then education. Education is huge and they promise the earth, I know a lot of it's down to funding...then you've got to keep kicking them up the backside to make sure that they're doing what they said they're going to. The specialist hospital were great at giving guidelines and on two separate outings they physically visited him at school to watch him in the class because they noticed that this child has made such a remarkable recovery, he should be further on. So they do a massive report telling the school what you need to do and they didn't follow it.”*  
(P1: 560-567)

*“So because of his behaviour he'd moved up to infant school, unbeknown to me they'd stopped him going out to out at break time and lunchtime. Because he was like a firework going off, he'd go out into the playground and he'd just explode. He needed to run around like a child with ADHD. So they stopped him going out and I didn't realise it, which means his behaviour then when he came home was that much worse. He effectively was locked in the same room all day. From half past eight in the morning till 3 o'clock in the afternoon. So of course by the time I went to collect him it was really hard to control him.”* (P1: 113-119)

### 3.1.3. Sub-theme 1c: Implications For The Family: The Knock On Effect

This subtheme highlights the challenges participants experienced within daily family life. This was a frequently occurring theme, mentioned by most participants.

The challenge of parenting was the most commonly mentioned and was most frequently highlighted within discussions about their child's behaviour. Difficulties around implementing boundaries or consequences were described in detail and,

for some parents, indicated a particularly challenging time where they felt they could no longer cope.

*“...his behaviour was just awful. I couldn't take him out. I couldn't trust him. ... He was just a terror. He was aggressive. He would hit out. And I just thought, oh no I just can't do it.” (P4: 51-53)*

*“...it's that guilt of actually, have you done this to your child? You need to put that aside and you need to be really strong and make sure that they're not walking all over you...Just being strong and setting boundaries and sticking to them is important because, yes ok they're not very well but actually you're going to have an awful time when they're older.” (P2: 552-557)*

Another instance, where a few parents felt unable to cope, was in relation to the difficulties of managing the health deterioration.

*“I got to the point where I thought I can't cope anymore because I've got a baby (sibling) that's not quite a year... I'm tryna drive and I'm looking in the back and I could see him turning blue in my mirror.” (P1: 162-165)*

As a consequence of these challenges, some parents went on to describe the ways their parenting style changed and adapted. Parents most frequently highlighted the need to be more protective, structured and organised in an attempt to manage the multiple behavioural and health demands.

*“Another really demanding side of it was keeping on top of it on a daily basis...Every day I would write down what meds he'd had, if he'd done anything quirky. What I needed to do, to chase it up. When the meds had to go up. When the meds had to go down. ...Organisationally it was demanding. It was like full time job.” (P8: 306-310)*

As well as adaptations to their parenting style, some parents talked about the necessity of contacting formal services to help support the management of their child's presentation. This was not always easy, as one mother notes the experience of guilt she experienced as a consequence of seeking respite:

*“And even with the respite, I would feel like a bad mother for sending her away. And after the six months she loved it and wouldn't want to come back. It's hard because it's not that you don't love, it's just you don't like them. It's so bloody hard” (P7: 502-504)*

As well as the challenges of parenting their child with LKS, this sub-theme highlighted the experiences of parenting their other children. A few parents described LKS as dominating their time, making it hard to engage with their other children in the way they would have liked, or had previously done.

*“It was pretty relentless. I found I didn't have the time to engage with my other son the way I had always done before. That I found that really really hard... I think it has had an impact him.” (P8: 216-219)*

As well as the experience of parenting, most parents also commented on the ways in which their daily lives had needed to change as a result of LKS coming into their lives. The financial demands of caring for a child with LKS was the most frequently reported experience, with many parents' employment status changing.

*“I know finances shouldn't come into it but it all impacts. It's got a knock on effect. We were spending endless days in hospital, which when your child's on the ward, you're not provided with a bed, you need to find money for the cafeteria” (P1: 316-318)*

A few parents also addressed their initial difficulty in planning ahead of time, highlighting that the severity of LKS meant that thinking about the future was too challenging. Due to the behavioural and health presentations, parents had to

remain focused on managing the current day and the needs their child presented with.

*“You couldn’t do anything or plan anything. Always had to plan being near a hospital. But as time went on and he got better, then the more confident we were and then we could start making plans.” (P1:534-536)*

The same few parents who expressed the difficulty in planning ahead also noted the negative impact LKS had on their social life. They described their daily life as being consumed with being a carer, leaving little space for socialising. This change seemed to be circular in nature, with the parents’ difficulty in finding someone who could take of their child, meaning they could not join friends. Consequently, the more they missed social events because of their child’s health, the more their friends withdrew. One parent also suggested that friends might withdraw because they do not want to hear about negative events.

*“I lost friends because of it. All our friends’ kids were healthy. All my friends were working going out for drinks on a Friday night. They could go out for a meal with friends. I had no one to look after X because my mom and dad refused because they were scared...So we didn’t have anyone to look after him. we didn’t have a social life.” (P1: 437-443)*

The impact of the changes to daily life were further described by some parents who shared how the family and their relationship with their partner had to adapt to the needs of the child with LKS. Although valuing each other, for a few there was an acknowledgement that different perspectives were often taken, which meant that their experience of having a child with LKS may have been slightly different.

*“There’s probably one other thing I’d add and that is the different ways couples deal with these things, you know because I think him and I deal with things very differently. In that I think he found the whole diagnosis really very difficult to digest because he was at work all the time. He [husband] didn’t see him [son] as much and he tended to be more black than white...Whereas I was with him more and being much*

*more proactive so I felt that I had a lot more positivity I suppose.” (P5: 470-478)*

### **3.2. Theme 2: Evolution of Family Roles and Ways of Coping Over Time**

This theme describes the changes that the participants experienced as a parent to a child with LKS and the subsequent strategies they drew from to cope . This was a dominant theme, mentioned by all participants and it related specifically to the changes experienced across the course of the disorder. The theme is comprised of five sub-themes: Advocacy: They're not Gods, Finding Strength and Power in Personal Resources, Acknowledging the Positive, Making Meaning and One Day at a Time

#### **3.2.1. Sub-theme 2a: Advocacy: They're not Gods**

The importance of taking an advocacy position for their child was seen to be a significant change for most parents and was the dominant sub-theme within this theme. The necessity of advocating for their child took various forms: the child's need for parental support; a perception that their health and/or academic needs were not being adequately addressed; and the desire to have the child regain their skills. Whilst taking an advocacy position, parents focused particularly on the need to be assertive with professionals. This sub-theme was expressed through personal examples or direct advice they would give to other parents.

*“This is my child and this is important to me. You need to help me. And if you can't help me, I'll find someone who can. I'm not normally like that. But I guess because he's mine. And I had it and I lost it. I wanted it back. That's basically it...I just kept thinking they are doctors but they're not Gods and they don't know everything.” (P4: 398-437)*

Some parents went on to acknowledge that an assertive advocate position can sometimes be difficult to take, especially in the context of already having to manage challenging experiences within the family home.

*“It was very draining and quite scary at times. I would come out completely shaking and often come home and burst into tears”  
(P8:345-346)*

*“You go into receptive mode even if you're a professional. Expect people around you to tell you things that they know to be true. And of course they don't all the time.” (P6: 72-73)*

In the context of advocacy and its importance, some parents also noted their observations of other parents who they believed did not take the same position. At times expressing this as a critique, parents shared their belief that unless they pushed professionals, their child would not receive optimal care.

*“I haven't spoken to a parent for a long time. I get a little bit frustrated with parents when they're not as passionate about it as I was and they don't seem to be pushing for their child. I feel like I want to do it for them because unless they push, they're not going to get heard.”  
(P4:445-448)*

### 3.2.2. Sub-theme 2b: Finding Strength and Power in Personal Resources

Most parents discussed the importance of drawing from the personal resources available to them to help cope with the challenges of raising a child with LKS. In this context I understood personal resources to mean both physical and metaphorical resources that empowered the parent to manage the demands of LKS.

For a few parents the importance of being able to access financial resources was viewed as a significant factor in helping their child to access appropriate support more quickly.

*“Also we were fortunate to have the money...so things like getting him into the speech and language school was because we had the help of an educational lawyer” (P5: 523-528)*

A few parents also drew on religious beliefs to provide an understanding of why LKS had entered their lives and to strengthen their belief they could cope. One parent expressed that this had not been easy and although she had doubted her faith at times, an acceptance that they would not have been given a child with LKS unless they could manage, was very important.

*“I'm a Catholic but I'm not a practising Catholic. But actually saying that, I have to believe that God gave me him for a reason because maybe I wouldn't have got through it...I can't see God's reasoning behind it, but I had to believe that at the time. God gave me him, because he thought I could cope. I had to keep telling myself that.” (P1: 481-485)*

As well as external resources, a few parents described the changes they had seen within themselves, enabling them to engage with professionals and their systems more efficiently. It seemed that the importance of being proactive not only provided practical benefits for the child, but also enabled the parents to stay focused and gave them a sense of purpose and importance.

*“But you see I was a fairly young naive stupid young mum, where all I knew was to care for my kids. Where I'd say I'm different now is that I know how to fight them. And then there was a battle... Yeah. because I was young and naive I didn't know what I was doing.” (P7: 122-126)*

*“...you look back and think, how did we go through all that, because we have files and files in our study...letters to schools...trying to fight for all these things. On one hand that's sort of a negative thing but on the other it gives you something to focus on which can be quite positive in a bizarre way.” (P5:517-521)*

For a few parents, this change seen within themselves was also understood to have subsequently shaped their work lives.

*“It has been a horrible road but that’s how I’ve ended up in this job. I went to volunteer for the ones [carers] that helped me...And it’s not the qualifications it’s because what I’ve done and been through.” (P7: 154-158)*

### 3.2.3. Sub-theme 2c: Acknowledging the positive

Whilst interviewing the parents, I noticed that the positive experiences encountered were often shared alongside discussions about more challenging times. The positive experiences mentioned included: the skills their child maintained; the regaining of skills towards the end of their child’s illness; and acknowledging the characteristics they valued within their child. Although the parents did not always explicitly identify the positives as a form of coping, it was evident throughout the interviews that these experiences were valued greatly and were shared as significant events in the course of their child’s illness, often representing when they were able to regain hope or a sense of normality.

*“I came round the bed and I put a little photo of his brother at the bottom and he pointed and said his name. I said to my husband, did you just here that?...Well that’s the first word he’d said in 18 months. Well I couldn’t believe it. I just cried and cried and cried.” (P4:183-190)*

*“So it’s not just the academic success, although that is really good to see because he always was a bright little spark. I feel like he hasn’t been cut off from his potential...But I really don’t know how one would have coped if things had been different. If our family would have stuck together. If it would have been different. I don’t know.” (P8: 577-580)*

*“He’s absolutely amazing. He’s so wise. I look at him and think I could learn so much from you. He’s only 20. He’s lovely. So lovely...You wouldn’t know, apart from the huge scar on his head that he had all these difficulties.” (P4: 420-424)*

*“For all the years I craved that little girl, that beautiful little girl I had, to now, I wouldn’t change. I’ve got grandkids now. She’s fantastic with*

*them. She's fantastic. Me and my daughter have set up our disco for disabled young adults 16 plus because X kept her love of music throughout her life...so we now do it every month and she dances her heart out and her boyfriend goes. She's got a better life." (P7: 409-415)*

A few parents also drew on real or hypothesised interactions with other children or families who were experiencing challenges perceived to be greater than theirs. These challenges included serious health needs, family structure (e.g. single parents) or different LKS presentations (e.g. significant behaviour changes).

*" You know what done it [helped to cope], being on the ward with a little girl with a tube coming out of her head and you think there is always someone worse than you. Always. And that's what we thought." (P3:332-334)*

*"We were told X's case of LKS was mild, we thought, this isn't mild for heaven's sake. Then we saw the other children who have been diagnosed not at seven or eight as X was, but at two. We realised how much more difficult it was for those children and those parents. I guess they have got that much less language to fall back on, you know. And so we were actually, we realise as you do when you are in these in these groups, networks, that there's always people worse off than yourself." (P6: 152-158)*

#### 3.2.4. Sub-theme 2d: Meaning Making

This subtheme draws on the value parents placed on developing a shared meaning and understanding of their child's presentation within the wider systems they were located. In turn parents were able to access support which promoted their ability to cope with the challenges experienced.

The positive support received from the parents' social network was mostly shared through examples of the practical support offered. Whether offering to look after their children or providing opportunities to gain knowledge about helpful

resources, the stories shared were presented with a lot of gratitude for their support.

*“Friends who lived close were wonderful and our really close friends took sign language courses with us so that we could continue with signing. And you know a couple of my very close girlfriends always offered to look after X...so our friends were amazing. They were just fantastic” (P5:328-332)*

The benefits of accessing support from the LKS community included the mutual understanding and guidance received.

*“We were flailing around. I think it was very important for us because we found parents who had the same experience, flailing around. As good as the medical care was, you needed some, a lot of it was about social, you know social wellbeing and how you sort of get the child forward.” (P6: 303-306)*

For a few parents, their own parents were felt to be a valuable source of support, presenting their non-judgmental stance and practical help as key factors.

*“My mum was really good. Because she was probably our only bit of support really...X loved my mom so she would sit and chill out. Most cases climbing up the walls. But most cases my mum would just allow her to do it. You know she understood that actually she wasn't well” (P2: 514-517)*

When parents described the benefits of accessing educational support for their child they highlighted the importance of staff having a clear understanding of the needs of their child (and sometimes of the parents) and witnessing academic and social development within their child.

*“But he [Head Teacher] was so brilliant because one day we were in a meeting and...he called her the Tasmanian devil on speed. I was ready*

*to punch him, but he wasn't lying. He was speaking the truth. I just sat there and thought you know, you're right. We got on so well." (P7: 184-186)*

*"In fact, the head mistress at the school, I'd read a paper that she'd done on working with a boy with LKS years before...I didn't realise she was the headmistress until I met her...So it all fell into place. He was in the right place" (P8: 138-141)*

When parents described the helpful or positive interactions with professionals within health and social care services, their examples highlighted times when the needs of their child or themselves were considered. The features of positive support included: helpful strategies; quick referrals to specialists; consistency; clear communication with parents and within the team; interaction with the child; feeling listened to; and showing an interest in parents' wellbeing.

*"...and really I just feel how amazing is it that she [doctor] was there from day one. When he was four and a half and had nothing. And she was there to sign him off. I mean she's known him throughout his whole childhood. And I just feel so lucky now because that consistency took the pressure off me." (P8: 411-417)*

*"So we went to see this doctor and was telling him about the journey so far. And he got on floor alone with X...he was the only doctor I remember actually interacting with this child." (P4:89-93)*

*"Looking back I do think that one of the things that stands out in my mind was meeting with one of the paediatricians at the hospital...a new consultant took over from him and he saw X and we had a conversation and he turned to me and he said, how are you mum? And that was the first time anybody, and we were some years into his diagnosis, had actually said to me how are things with you...it really horrified me at how emotional it made me feel...to have somebody*

*actually ask you how I felt both overwhelmed me with the question but was just so pleased someone at last thought, how about you.” (P5: 160-171)*

### 3.2.5. Sub-theme 2e: Taking One Day At A Time

Most parents talked about the importance of time. Although ‘accessing helpful resources’ and ‘acknowledging the positives’ were significant themes in people’s stories, they were mostly presented as reflections made in hindsight. The importance of taking every day as it came was mentioned as a direct recommendation by a few parents, as they addressed the overwhelming nature of their experiences.

*“ And how we got through it. It was a case of just took every day as it came. An awful lot of heart ache and an awful lot of tears.” (P1: 468-469)*

*“It was existing wasn’t it? Making sure she had a bed and the other kids went to school. It was just existing. You can sit and talk about it, but you’ve just got to get on and live it.” (P7: 463-464)*

Some parents expressed surprise at how their memories of the challenging years and their earlier feelings of distress had faded over time. Once their child regained their skills, their lives had moved on.

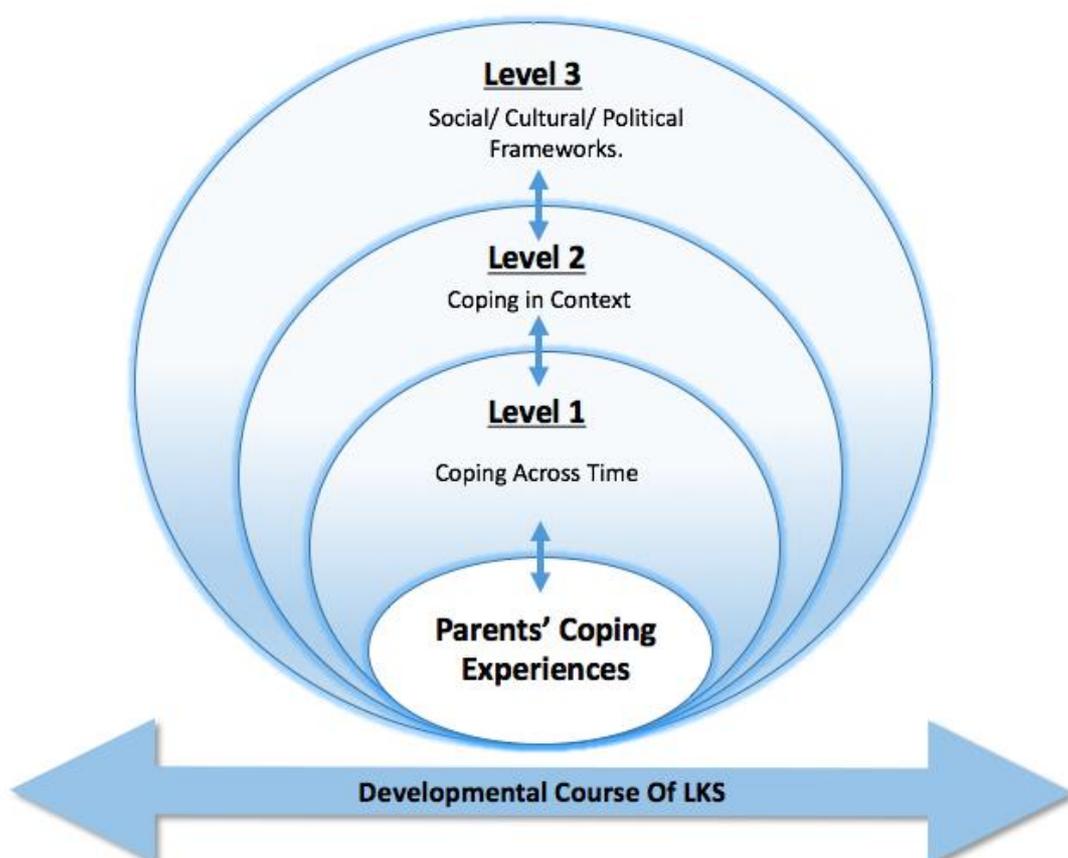
*“When on occasions other mums have rung me up. Maybe just had a diagnosis or they are into early parts of the diagnosis or meds. I sort of realise how much I’ve forgotten. It’s not that I have, I haven’t forgotten. I can remember. It takes me straight back to that sort of. Just the intensity of the anguish...and it doesn’t feel like that anymore. It really doesn’t. I’m optimistic for the future really.” (P8: 425-432)*

## 4. CHAPTER FOUR: DISCUSSION

This chapter begins with an exploration of the themes derived from the analysis in relation to the research questions and relevant literature. I then consider the wider implications of the findings for psychologists practising at a clinical, service and research level. The chapter concludes with a reflective account of the research process and a critical review of the methodological limitations.

### 4.1. Discussion of Themes

#### 4.1.1. Psychological Factors/Models that Influence the Parents' Coping Experiences



*Figure 1. Psychological Factors/Models that Influence the Parents' Coping Experiences*

Figure 1. presents the key psychological factors and models I am utilising to conceptualise and discuss the themes revealed within the analysis. Informed by the work of Kazak and Nachman (1991), I have been inspired to draw from theories that focus on systems and their importance to understanding families' experiences of LKS and how they cope. These models include Bronfenbrenner's ecological systems model (1979), CMM (Cronen, Pearce, & Changsheng, 1980) and a biopsychosocial perspective. Overall I aim to demonstrate that the parents' experience of LKS and they way they cope is the result of an interplay between different micro-to-macro-level influences.

Level 3 represents the overarching factors which indirectly affect the parents' experiences through their influence on level 1 and 2. Influential developmental theories provide a framework to navigate typical child development and thus promote societal and parental expectations of children's behaviour as well as the expected role of parents. A cultural framework of knowledge can also shape dominant discourses about parenting, as well as influence understandings of health.

Level 2 highlights how these overarching frameworks can interact with the parents' daily experience within the family and society (Carter & McGoldrick, 1999). Through societal norms and dominant frameworks of knowledge, family roles and expectations are created. Furthermore, the values held from the dominant framework of knowledge will shape expectations of who will be best able to support them and how (e.g. community leader, professionals).

Level 1 represents the psychological factors that are understood to directly affect the parents' coping experiences. Concepts such as the stages of loss and coping (Sen & Yurtsever, 2007), Maslow's hierarchy of needs (1943), 'Care-giver Burden' (Jordan & Linden, 2013) and 'Chronic Sorrow' (Copley & Bodensteiner, 1987) can also be seen as dynamic features, influenced by the factors at levels 2 and 3. All levels are further contextualised by the developmental stage of LKS.

For example, the child's presentation during the initial onset or side-effects of treatment are crucial considerations when understanding the parents' coping experiences.

#### 4.1.2. Research Questions

Whilst discussing the themes presented within the analysis, the two research questions outlined at the beginning of the study will be addressed. and discussed in the context of both themes; 'Challenges to Coping' and 'Evolution Of Family Roles and Ways of Coping Over Time' .

This theme address the many challenges parents experienced as a consequence of being a parent to a child with LKS. These challenges can be understood to impact a parent's belief they can cope, and set the context for the ways in which families adapt and manage to cope over time.

Parents witnessing their child's health deteriorate at the initial onset of the disorder was highly significant. This period of time was associated with descriptions of shock and distress, compounded by the challenge of the disorder being so rare (NIH, 2015). The earlier developmental course of the disorder presented significant parental challenges.

Parents started all interviews with detailed accounts of the initial deterioration of their child's behaviour, skills and health within the context of typical development. Using emotive language, parents expressed the unbelievable and shocking nature of this change. Their subsequent experiences of anxiety, fear and distress correspond to the 'primary reactions' described within Sen and Yurtsever's (2007) parental response categories. As outlined within this and other models of parental adjustment, the experience of loss was pertinent and seemingly underpinned the emotional reactions (Anderegg et al., 1992; Smith et al., 2015). The parent was left with a child that no longer resembled their own and consequently, the severe distress felt can be compared to experiences of grief for the child that metaphorically was 'lost' or 'died' to LKS (Hewson, 1997).

Over the course of the illness, parents went on to highlight the difficulty of watching their child re-gain and lose skills again. Additionally, for some parents, their child's lack of observable seizures and the subsequent 'invisibility' of LKS exacerbated this challenge. Drawing from the theorists who have conceptualised a staged process of parental adaption and coping, the necessary and final position of 'resolution of loss' may be hard to achieve because of the inconsistency of their child's presentation and the possible hope of recovery (Sen & Yurtsever, 2007). Instead, parents' experiences may better align with the concept of 'chronic- sorrow', where feelings of loss and grief fluctuate between happiness, as parents experience different presentations of their child along the course of the illness (Hewson, 1997).

The significance of these experiences can be further understood within the wider societal context, where particular norms of child development are expected and valued. The unexpected nature of their loss was powerfully communicated during the interviews and emphasised the challenge of witnessing their child's presentation deviate from what was anticipated.

Dominant developmental and societal expectations can also be seen to underlie the concern some parents expressed regarding the emotional wellbeing of their child as a result of LKS. The discomfort in watching their child encounter social exclusion was particularly upsetting for some parents, suggesting that significant value was placed on these interactions. Although not discussed in detail within the interviewees, it could be argued that as well as parents holding knowledge that peer interactions are a valuable developmental milestone, they may also represent societal acceptance. In relation to dominant western values on social independence and economic success, and the family's own expected life cycle stages, observing their child's exclusion may raise concerns about their child's ability to achieve these anticipated norms and the consequential parenting they may need to provide (Carter & McGoldrick, 1999; Waldboth et al., 2016).

As briefly discussed in the work of Cockerell et al. (2011), the rarity of LKS seemed to further compound the distress of witnessing their child's deterioration and created a barrier to accessing support from the health and social care

systems. Drawing from literature highlighting the significance of a 'diagnostic gap', the delay between initial symptoms and definitive diagnosis led the parents to experience a multitude of challenges (Berglund, 2014).

Paucity of relevant medical information about the disorder perhaps made it difficult for the parent to adapt and gain knowledge that might serve to contain worries about the future outcomes of their child (Berglund, 2014; Pelentsov et al., 2016). Furthermore, the lack of information about the cause of LKS, experiences of inconclusive medical investigations, dismissal of parents' concerns and initial misdiagnoses, may have led parents to question their actions and their control over the outcome of events (Ellis, 1989). Consequently, feelings of anger can turn inward, magnifying a parents' upset (Findler et al., 2016)

The rarity of the disorder also explicitly highlighted the consequential challenges of interacting with professionals within health and education settings. Parents frequently described the battles and hurdles of accessing appropriate support for their child. As seen within research on other rare disorders, an overarching feature of their experience was related to their frustrations about the perceived lack of knowledge the professionals held about the disorder (Berglund, 2014; Grut & Kvam, 2013; Fletcher, Flood, & Hare, 2016). Parents described their experience of being mediators between specialist and local teams, taking responsibility for treatment or education intervention guidelines being followed. Furthermore, their stories also presented a lack of attunement with professionals, who seemingly rarely considered the emotional impact of their experience.

Although this lack of attunement could be considered a repercussion of the professionals' lack of consideration for the parent, a few parents shed light on the complexity of the relationship. Two parents noted a 'false' presentation of strength and coping given in meetings, perhaps making it harder for professionals to identify where support may be needed. As discussed previously, the interaction between parent and medical professionals may be bound by assumed roles (Budysh et al., 2013). Neither parents nor professionals may consider medical appointments as an opportunity to discuss the parents' personal experiences, therefore the parents' needs are not addressed.

These challenges may also be best understood via a broader cultural and societal framework. Dominant western narratives present health and treatment as a universal, objective experience, with medical professionals holding particular status and expected behaviours (Budysh, Helms, & Schultz, 2012). However, through the parents' unhelpful experiences with professionals they gained insight into the subjective nature of health and recovery. No longer viewed as competent experts who provide all answers, the traditional role structure between patient and professional is challenged (Budysh, et al., 2012). This may be further affected by the societal shift towards becoming active participants in care, where they are encouraged to become involved in health policy and health care services (Coulter, 1999).

In the context of Informal support, interactions with the community and family were shared. As seen within wider literature (Ludlow et al., 2012; Pelentsov et al., 2016), the research identified an underlying frustration and sadness that others lacked understanding of LKS. In the context of family and friends, the impact of their child's social behaviour played a significant part, often leaving parents feeling judged and socially excluded. Within the context of other parents of children with LKS, the difficulty in being able to relate, due to the differences in health and social factors (e.g. gender of their child, severity of disruptive behaviour and stage of illness), led to unproductive interactions.

Participants also drew attention to the many ways in which LKS impacted the family. The participants' stories relating to the challenges of parenting, were most frequently discussed in the context of behaviour. As seen within ABI and ASD literature, disruptive and unpredictable behaviour was related to experiences of 'carer-burden' (Brown et al., 2013; Kim et al., 2016). This concept is thought to encapsulate daily life feeling less 'normal' and parents having less confidence in managing their child's condition (Kim et al., 2016). Parents described how overwhelmed and unable to cope they felt at times.

This lack of confidence in managing their child's presentation could be responsible for the parental accounts which reported the need to contact formal

services for support. Sen and Yurtsever (2007) highlight this need to access support within their 'tertiary reactions'. Outlined as an ongoing phase, parents are thought to develop an understanding of their unexpected situation, accept that changes need to be made in their lives and begin to utilise help from professionals. Although discussed in more detail within the next theme, this engagement with services was not simple and was often described as challenging.

The challenges of parenting and engaging with support could be linked to dominant societal and family discourses about the role of parents. A belief that parents, predominantly mothers, hold great responsibility for their child's outcomes and behavioural presentation may underpin the distress and stigma experienced (Francis, 2012; Grey, 2002; Leskošek, 2011). Additionally, parents' discomfort in requesting external support can be understood as a reluctance to publicly acknowledge their struggle or to expose themselves to stigmatising beliefs about the limitations to their own parenting skills (Francis, 2012; Grey, 2002).

As described in most parenting literature, caring for child with health needs is a multi-faceted experience, which is not limited to the experience of parenting the one child (DePape & Lindsay, 2015). Drawing from a systemic perspective and its assumptions of interdependence within the family, it is perhaps unsurprising that parents within the current study also shared difficulties whilst parenting their other children. Although not mentioned by many parents, this experience is acknowledged within wider literature, with parents wanting to compensate and reach out to their other children because they spent more time with the child diagnosed with a disorder (DePape & Lindsay, 2015). It could be that siblings were not mentioned more frequently because it felt somewhat shameful to discuss neglect of one child in the context of discussing advocacy and exceptional care given to another.

Another difficult aspect of family daily life was the social implications for parents. Predominantly discussed in the context of their child's disruptive behaviour, parents described no longer socialising with friends. The lack of understanding

and negative judgements from others were particularly difficult for parents and mirror the parental reports within the more widely researched area of ASD (Ludlow et al., 2012; DePape & Lindsay, 2015). Negative critiques resulted in the parent feeling judged and blamed, sometimes preferring not to go out in public with their child. This is a valuable insight, as it presents how dominant societal expectations about parental responsibility and child norms, creates social stigma which further leads to parental negative self-judgment (Grey, 2002; Wong et al., 2016; Jordan & Linden 2013).

Finally, within this theme, the financial demands and concerns arising as a repercussion of their child experiencing a rare condition echoes the findings of wider literature on this topic (Pelentsov et al., 2016; Zurynski, et al., 2008). What was also interesting about these accounts is that this topic was shared with a sense of shame, presenting an unease with highlighting financial concerns in the context of wider worries about their child's health. Again, dominant discourses around caring for a child may be influential. The image of altruism within the parents' role perhaps marginalises the more complex narratives about the needs of the parent (Francis, 2012). Drawing on the work of Maslow's hierarchy of needs (1943), this is significant because if basic needs are not met, negative psychological consequences may arise.

#### 4.1.3. Theme 2: Evolution Of the Family Roles and Ways of Coping Over Time

Moving away from a previously dominant focus in the literature, which simply highlights the 'negative' experiences parents of children with childhood disorders, there has been increased interest in the ways in which families cope (Atkin & Wagar, 2000; Rolland & Walsh, 2006; Knafl, Deatrck, Knafl, Gallo, Grey, & Dixon, 2013; Kazak & Nachman, 1991; Cipolletta et al., 2015). This theme aims to give a detailed account from the parents' perspective of what it means to cope with having a child with LKS. Drawing on the importance of; advocacy, the use of personal resources, acknowledging positive experiences, developing meaningful collaborative understandings and taking one day at a time, this theme describes the ways in which parents coped with the challenging experiences of having a

child diagnosed with LKS over time. It was seen that parents' adjustment and ability to cope was dynamic and affected by their child's presentation and changes to the family's needs.

In support of the findings by Cockerell et al. (2011) and Lemard-Reid's (2014) work, the importance of taking an advocacy position for their child was highlighted by most parents interviewed. The dominance of this sub-theme suggested that parents had not anticipated the level of advocacy necessary, especially alongside the perceived responsibility for their child's health outcomes. In the context of the devastating loss and lack of knowledge held by professionals, experienced by parents at the early stages of the illness, the importance of advocating as a way of coping seemed linked to the importance of gaining control and power. Critiques of other parents' choices reinforced the importance of advocacy. Disappointment and frustration regarding flawed professionals moved parents to value and articulate their own knowledge and thereby become more empowered.

In line with the work of Delleve et al. (2006), this emphasis on taking an active style of coping with the engagement of practical acts, seemed to enable the parent to feel that they were 'doing something' and taking control. The fight for appropriate educational or medical support seemingly gave parents the opportunity to protect their child in a manner they had been unable to do against LKS. The importance of expressing and acting on this perceived responsibility to protect their child can be linked to wider societal and family roles valued by the parent.

Thoughtfully, parents acknowledged that this position was sometimes challenging to take. The preferred medical model framework of knowledge naturally positions professionals in a more dominant position and could be seen to affect the parents' perceptions of themselves, with occasional self-descriptions as 'naïve' and 'stupid'. Two parents, medical professionals themselves, explicitly named their own experience of doubting their well-informed medical opinion as they interacted as parent to professional, rather than professional to professional. Within this context, it is perhaps not surprising that most parents felt this was a

significant experience for them, as it required an active resistance against a submissive position typically held, or maybe societally expected of them (Budysh, et al., 2012).

The parents' ability to draw from their personal resources was seen to be a significant strategy for all parents. The importance of financial resources seemed crucial for a few. However, the topic of money was shared with some hesitancy. The topic was consequently discussed with the acknowledgment that most parents' lack of access to this resource was fundamentally unfair. These findings support features of Lemard-Reid's (2014) parental reports, which propose that socioeconomic factors are a significant element of the parent experience of LKS and are perceived to affect the speed of diagnosis and support available. Parents who have access to financial resources could be seen to develop a different power relationship with the professional, as they are not required to accept the knowledge provided (Benzeval et al., 2014) but can access alternative resources until they find support that is believed to best suit their and their child's needs.

The development of a strong identity continued throughout the interviews as some parents explicitly described the personal changes they had seen, including increased knowledge and a confidence to express themselves. For a few, this change was felt to have a direct impact on their ability to engage with professionals and navigate systems more effectively. These narratives support the less widely reported literature that caring for a child with additional needs can bring about a positive parental identity of resilience and growth (Nicholas et al., 2015; DePape & Lindsay, 2015).

For a few parents, the importance of looking to a religious explanation also seemed to help them cope. Exemplifying the impact of differing frameworks of knowledge, religious beliefs were a source of comfort, and reassurance of the ability to cope (Atkin & Wagar, 2000; Danseco, 1997; Kazak & Nachman, 1991).

The description of positive experiences encountered and reflection on real or hypothesised interactions with other children or families who are perceived to experience challenges greater than theirs, seemed to give the parent strength,

resilience and motivation to continue supporting their child. Making downward comparisons in the belief that things could be worse, put into the focus the resources they had available and elevated their self-regard (Wood, Taylor, Lichtman, 1985).

Although mentioned less frequently, the helpful experiences of accessing support from formal provision highlighted the benefits of professionals taking a non-judgemental position. From this position, parents experienced validation of their concerns and attunement through a collaborative understanding of both their and their child's needs. As seen in literature on rare disorders, this non-judgemental stance led to parents feeling less frustrated with professionals' lack of knowledge (Kirk et al., 2015). Instead parents valued professionals who were transparent about the limited knowledge they held and were able to position themselves alongside the parent, helping them seek further expertise to support their child. These experiences are particularly pertinent in the context of stigma and power highlighted previously and suggest that parents' interactions with support can be significantly affected by the approach of professionals (Francis, 2012; Grey, 2002).

As discussed within Pelentsov et al., (2016), helpful interactions within informal contexts predominantly focused on the importance of practical support. Either in the form of child care or advice and guidance, the benefits of understanding and non-judgemental peer support led to the consequential belief they were not alone. This experience was felt to be invaluable in the context of the challenges faced within the dominant formal setting on which they were dependant.

Finally, parents talked about the importance of taking every day as it comes. It seemed that thoughts of the past and predictions for the future were overwhelming, therefore a focus on the daily challenges was experienced as relatively more manageable. This reluctance to recollect the past seemed to continue into the current day, with many parents commenting on their surprise at the memories that were arising during the interview. This experienced could be conceptualised through a process of 'active forgetting', where not remembering

traumatic memories enabled them better function in day to day life without being disturbed by past distress (Anderson, 2001).

Overall the findings support the critiques of staged models to coping. It was clear that acknowledging the individuality of parents' experiences of coping was crucial. As highlighted within Kazak and Nachman's\_(1991) research on parental coping, parents' adjustment and ability to cope was seen to be dynamic and based on the many ecological systems in which parents, their child and childhood disorder is experienced within. The process of coping was interactive and also changed over time. As parental knowledge grew, their ability to draw from more proactive coping strategies and personal resources, increased. In turn, interactions with professionals and others began to be experienced as more helpful. Collaborative and helpful joint meaning making allowed for the parent to feel understood, listened to and therefore more receptive to the input offered.

## **4.2. Implications and Recommendations**

The findings of the current study have provided a novel insight into the experiences parents consider significant when having a child diagnosed with LKS. As this research used a thematic methodology with 8 participants, generalisations are difficult. However, the parents' interviews not only provide a rich picture of their personal experiences of parenting a child with LKS, but give insight into their experiences of engaging with education, health and social systems. Based on a belief that parents are a source of support and knowledge in promoting child health and wellbeing outcomes, the direct and indirect implications will now be discussed within the context of clinical, service and research level recommendations.

### **4.2.1. Clinical Level**

The findings highlighted that the parental experience has a significant personal impact on parents, including how they manage to cope. As understood within wider paediatric literature, parental wellbeing can have the potential to positively affect their child's health and wellbeing outcomes through pathways such as

increased attentive caregiving (Feeg et al 2016; Brilli et al., 2014; Kuhlthau et al., 2011). Consequently, it may be useful to routinely offer psychological support as part of their child's treatment plan. Psychologists working on an individual level may find it beneficial to integrate psychological and health models to allow a more comprehensive formulation of the multitude of challenges associated with LKS. For example, utilising the ICF-CY framework within the context of Bronfenbrenner's social-ecological systems perspective can help conceptualise the intersection of particular health and social factors the parent is experiencing. These factors might include; the stage of illness and their child's subsequent presentation; the child's stage of development and anticipated parental role; their preferred framework of knowledge from which they make sense of their experience; and their expectations and experience of accessing support.

Additionally, the ways in which some parents spoke of LKS and their position of strength lends itself to narrative therapeutic interventions (Morgan, 2000). Adopting a narrative model could build on the already naturally occurring externalising conversations and help the family to develop and strengthen preferred stories. In turn this can promote their belief they can cope.

When considering the potential importance of psychological support, it also feels important to consider the timing of therapeutic interventions. From the stories shared it may not be viable or desired to access support whilst experiencing the particularly challenging times of parenting. Therefore, the offer of support should be available at any time during the course of the illness.

This consideration prompts an acknowledgment of the access to peer support and its potential role in mediating parents' ability to cope (Kazak & Nachman, 1991; Wallander et al., 1989). Parents of children with LKS describe social isolation and strains on family relationships. Moving beyond the individual, it seems crucial to provide opportunities to receive peer support through parent networks. From the stories shared, parents found this support particularly beneficial when it provided practical guidance. Gaining emotional support from others within the LKS community was presented as more challenging because of the recognition that all parents experience of LKS is slightly different.

In response to this challenge, it is important to develop an opportunity where a wide network of people at varying stages can be accessed. Due to the rarity of the disorder, the broad distribution of the population around the country may make an online forum useful. This could be a peer-led site that promotes the distribution of practical advice and strategies, as well as giving opportunities for people to make contact with others who can relate to the particular and varied aspects of their experience and therefore provide the desired support.

#### 4.2.2. Service Level

Although distress among parents of children with health disorders is well acknowledged, the findings of this study suggest that particular consideration be given to the wellbeing of parents whose child's disorder is also rare (Atkin & Wagar, 2000). Although parents may present to be 'coping' at routine appointments with medical professionals, it should be acknowledged that these parents may not be able to gain the reassurance or containment available to parents of children with more well-known disorders. Consequently, a pathway to accessing the support outlined above should be routinely incorporated into treatment offered.

In relation to the challenging dynamics described between parents and professionals, health and social care teams should try and ensure parents experience interactions where they are heard and their experiences and feelings valued. It may be useful to invite parents to multidisciplinary team meetings or convene a network meeting. Drawing on the work of Anderson and Johnson (2010, p.113), network meetings can provide a space where large complex teams come together with the family to discuss queries and issues. This may be particularly relevant for parents who experience disparities between the advice given and interventions offered by their local and specialist teams. Furthermore, it may also be beneficial for the psychology team to offer consultation or advice to their professional health care colleagues, to support them with the insecurity and lack of knowledge on LKS. This would aim to promote helpful transparency by professionals and remove the potential defensiveness that can get in the way of

professionals acknowledging that parents may hold valuable information that could lead to positive child wellbeing outcomes (Budysh et al., 2012).

In the context of the particularly challenging experiences with schools, it is important to note that parents who received support from the hospital to educate the staff on LKS had found it to be a very helpful experience. As described by Abrams (2014), the communication between teachers and parents can strengthen or weaken the bond between school and home, leading to beneficial outcomes for the child. Therefore, psychoeducation on LKS should be offered routinely to the child's school to promote an understanding of the challenges faced and beneficial interventions. This promotion would not need to be facilitated exclusively by a psychologist or other health care professional, but could be facilitated by a parent who has relevant knowledge on LKS presentation and management strategies.

#### 4.2.3. Research Level

##### 4.2.3.1. *Dissemination*

The lack of knowledge about LKS on a wider level led the parents within this study to struggle with local health authorities and the educational system, in order to obtain the correct diagnosis and receive adequate help. It is therefore important that an awareness of LKS is promoted within medical, educational and speech and language settings to reduce the burden experienced by parents (Berglund, 2014). Findings from the current study could be disseminated at conferences for paediatric health care providers as well as published in healthcare journals.

Although, at the time of writing this study, there were no LKS peer-parent groups known to myself, these research findings can be disseminated to any future groups developed. The findings may serve as a tool for parents to gain insight and knowledge about the potential challenges faced and the resources others found helpful. I will begin by distributing the report to the parents who were interviewed, with a recommendation to share with other people they think may be interested.

At the time of writing this study, I had shared my preliminary findings with a developmental epilepsy team within a specialist London hospital. Seeing many children with LKS from across the country, they were keen to hear about the experiences of parents and, in particular, how the findings could be used to help their team facilitate a parent group. I will therefore be sharing the importance of addressing the complexity of the wider parental experience alongside their specific experiences of accessing formal and informal support. In turn this will continue to promote the important principle that, if parents' experiences are listened to and valued, helpful changes can be made to service intervention, evaluation and practice.

#### *4.2.3.2. Future Research*

Employing a thematic analysis methodology, enabled novel insights into what it is like to be a parent of a child diagnosed with LKS. However, it is important to note that parents provided a retrospective account of their experiences. Future research with parents whose children have recently received a diagnosis may highlight different, unexpected insights that the parents within the current research were not able to access. Additionally, these findings may have greater applicability for current clinical practice and service delivery.

A wide perspective was taken within this study, utilising a systemic framework. Taking this approach has its advantages and disadvantages. An advantage is that it enabled the experience of LKS to be conceptualised within multiple contexts, as recommended within research on coping by Kazak and Nachman, (1991). Consequently LKS was seen not just to impact the child and was instead seen to have a dynamic and complex impact on the family. A disadvantage of considering the multiple layers of context, is that it may obscure a more fine grained analysis of family coping dynamics. For example, a narrower focus may have led to an exploration of the differences in male and female parental reactions. In the context of wider literature and the current research, the majority of participants were mothers. An explicit focus on fathers' experiences may highlight a more nuanced understanding of their experience of LKS and how support could be provided. Furthermore, the importance of the sibling experience could also be explored. This would give further depth to our understanding of how

LKS impacts the wider family and consequently lead to recommendations on the support that could be beneficial. From the stories of discomfort and guilt around the parenting of their other children within the current research, a focus on the wellbeing of siblings could also promote the wellbeing of parents.

### **4.3. Critical Review: Evaluation of Current Research**

Yardley (2000) highlights that in the context of health psychology research, it is necessary to consider how the value of a piece of qualitative research should be assessed. This sections aims to present an evaluation of the current research through the consideration of four key criteria: quality, validity; methodological limitations and reflexivity.

#### **4.3.1. Quality and Validity**

Yardley (2000) suggests four principles be adopted to ensure validity within health psychology research: 1) sensitivity to context 2) commitment and rigour 3) transparency and coherence and 4) impact and importance. I will use this framework to discuss and address each element of quality and validity in turn.

##### **4.3.1.1. *Sensitivity to Context***

‘Sensitivity to context’ requires the researcher to attend to the theory, relevant literature, sociocultural settings, perspectives of participants and ethical considerations. To ensure sensitivity to context was upheld, I conducted a search of the existing literature investigating the experience of parents within the paediatric health context. This also ensured I had addressed the relevant theory. Additionally I sought ethical approval and attended a ethics committee meeting. This process forefronted the ethical considerations of the research, encouraging me to explicitly consider the potential impact on participants and how this will be addressed. Finally, I conducted in depth interviews and used verbatim quotes to ensure the perspective of the participant underpinned my analysis and conclusions.

#### *4.3.1.2. Commitment and Rigour*

'Commitment and rigour' address the need for researchers to have an in-depth engagement with the topic, express their competency in the methodological skills being utilised and to conduct a thorough process of data collection and analysis. Thematic analysis requires an in-depth engagement with the data, therefore I have described each stage of the analysis process within the methods chapter. To ensure competency I have utilised relevant teaching at university and attended peer-led seminar sessions to develop and improve my analytic skills. Furthermore, each of the themes and sub-themes were reviewed by my academic supervisor and field supervisor leading to discussions and revisions which prompted further learning.

#### *4.3.1.3. Transparency and Coherence*

'Transparency and coherence' highlights the need to have clarity when describing the data, to be transparent in methods and data presentation, to make links between method and presentation and to demonstrate reflexivity throughout this process. I have attempted to present the data and findings of the current study clearly, enabling the reader to gain a coherent understanding of what was expressed by the participants. I have provided detailed examples of each stage of the analysis in the appendices to ensure transparency and located the findings in context to the current literature. Reflexivity will be addressed later in this discussion chapter.

#### *4.3.1.4. Impact and Importance*

'Impact and Importance' recognises the theoretical, practical and socio-cultural implications of the research. This has been outlined in the first part of this chapter, highlighting how the findings of the current study have provided a novel insight into the parents' experiences of LKS. This was discussed in the context of direct and indirect implications at a clinical, service and research level. Additionally, I address how I plan to disseminate the research, highlighting the importance of initially sharing the findings with the participants.

#### 4.3.2. Methodological Limitations

Qualitative research is aimed at gaining a rich understanding of people's experiences and perspectives in the context of their social worlds (Braun & Clarke, 2006). Although the current study aimed to meet the criteria outlined by Yardley (2011), it is necessary to further evaluate the limitations to the methodological approach used. As highlighted by Willig (2008), it is important to address the critiques of qualitative research which include generalisation of the sample researcher bias and transparency.

##### 4.3.2.1. *Recruitment and Sample*

A purposive homogenous sample was recruited with the aim to develop an understanding of the parental experience of having a child with LKS. Throughout the course of conducting the current research I became aware of different limitations affecting this aim. This can be best understood through the discussion of my recruitment process and subsequent sample of participants I interviewed.

Despite there being an agreed diagnostic criteria for LKS (Hirsch et al., 2006), Stefanatos, Kinsbourne and Wasserstein (2002) highlight the complexity of this disorder and the subsequent variability in clinical expression. This variation can be seen within the current sample and in the range of different experiences parents encountered. This variation may make generalisations from the current findings challenging, as parents from the wider LKS population may face experiences different to the current parents.

In regards to the generalisability of findings to parents and professionals recently encountering LKS, all parents were selected using an inclusion and exclusion criteria which selected parents whose children were no longer in the 'active phase' of LKS and whose diagnosis was not recent. This criteria was chosen on an assumption that parents with a historical diagnosis would have a broad range of experiences from which to draw. However, the impact of this criteria is that professionals' knowledge and understanding of LKS may have changed and developed since the time of the current participants' experience of diagnosis. Additionally, clinical practice and professionals' attitudes towards working with children and their families may have changed. If so, this could have had a

significant impact on how LKS was addressed with parents within health and educational systems.

It was also noted, when conducting the interviews, that nearly all parents reported that their child had recovered from LKS with no significant difficulties. As presented within medical literature, this is not always the case and some children experience lifelong language difficulties (Metz-Lutz & Filippini, 2006; Caraballo et al., 2014).

*“...it did occur to me that I don't know how I would feel about any of it if the outcome had been different. I actually don't know if I would have coped... if I felt I'd lost X forever. I think it's because he's come out the other side I feel all right about it. I'm aware there are people who haven't.” (P8:556-560)*

This observation presents questions about the nature of research engagement. In the context of child health outcomes and parental responsibility described within the current study, were parents more willing to participate and discuss their experiences because their outcome was positive? Did the parents who decided not to be interviewed have experiences that prompted a belief they had not 'succeeded' and their experiences were not significant enough to be heard or learned from? Consequently, this bias within the sample should be acknowledged if attempting to make generalisations of the findings to the wider population whose children are thought not have made a 'full' recovery from LKS.

Further consideration for the sample interviewed leads to a discussion about demographic features. In light of wider parental reports, fathers were underrepresented within the current study (Phares, Lopez, Fields, Kamboukos, & Duhig, 2005). Therefore this study cannot claim to have presented an exploration into the potentially different experiences of parents of different genders.

Furthermore, although I have contextualised the data within the methodology chapter by providing basic demographic details, I did not report in-depth participant descriptions due to confidentiality (Sanders, Kitzinger & Kitzinger,

2015). This omission may also have served as a barrier to novel insights and understandings of the more idiosyncratic factors as well as patterns across families which will have affected their experiences and informed their perspectives.

Finally, it is important to note that the current study interviewed eight participants. Although Baker and Edwards (2012) stated that there was no exact number of participants required within a research study, I acknowledge that a larger sample size may have provided a more nuanced understanding of the themes identified. However, due to the rarity of the disorder, finding more participants would have been particularly challenging.

#### *4.3.2.2. Interviews and Analysis*

Individual interviews offer a vast amount of information. Rapley (2001) argues that interviews are inherently social interactions. The data gathered is dependent on a specific interaction and, consequently, the knowledge produced in interviews is co-constructed between the researcher and participant (Larkin, Watts, & Clifton, 2006). Stevens et al. (2009) go on to highlight the importance of creating a cooperative approach between the researcher and participant within the context of families caring for children with health disorders. I incorporated this into my approach through giving particular consideration to the sensitive and emotional content of the interviews, and through valuing engagement and interview preparation.

As I recruited through a London specialist paediatric hospital, where most families have received care, I am aware that my presence may have represented this establishment. This hospital has an international reputation as a provider of expert knowledge and high quality care. Although I was not a member of staff, sharing less positive views about the establishment may have been difficult for some parents. Conversely, due to my perceived status or position within the hospital, some parents may have felt this was an opportunity to 'feedback' and have the service bear witness to the challenges they faced as a consequence of the care received.

In an attempt to reduce the potential power imbalances occurring due to my perceived professional position at the hospital or, more generally, as a researcher, I was flexible regarding the times and locations of the interviews. All participants opted to be interviewed in their homes and many commented on the length of my journey. It seemed that for many parents, the gesture of travelling to their home embodied my desire to learn from them and conceptualise their experiences as valuable knowledge. In turn this promoted participants to feel safe, comfortable and discuss subjects openly. Cotterill (1992) goes on to suggest repeated interviews could have also prompted a deeper level of trust and further disclosure of rich data.

Finally, I acknowledge that thematic analysis and presentation of these findings is just one interpretation. Influenced by both the researcher and the research questions, it could be argued that a different methodological approach would have produced different findings. For example a narrative approach would elicit a more detailed understanding of how they make sense of their experiences and how these are constructed within the social and political environment in which they exist.

#### **4.4. Reflexivity**

Engaging in reflexivity is understood to be crucial in conducting rigorous and valid qualitative research, as no single objective truth or reality is assumed (Harper, 2012; White, 1992). Consequently, I aim to reflect and acknowledge how I have contributed to the construction of meaning and therefore the research itself. Throughout this process I have questioned how my existing knowledge, prior research interests, values and experience of the research topic have been influential during the course of conducting this research (Dibley, 2011; Ely, Vinz, Downing & Anzul, 1997).

##### **4.4.1. Reflexivity In The Design Of The Study**

Whilst exploring the literature and developing my research questions I noticed myself drawing on my previous academic and clinical experience. Prior to

commencing Clinical Psychology training, I had worked as a teaching assistant and conducted research in schools with parents whose children had health conditions. Initially I was apprehensive that the current study would not bring to light any novel findings and would instead replicate the wealth of knowledge presented on more common health conditions. However, whilst conducting the literature review, I noticed that my understanding of research had changed since completing my Masters degree. Through increased clinical experience I developed a more sophisticated understanding of people's experiences and how individual factors could intersect to create a unique and powerful narrative. In the context of LKS, I became intrigued with the interplay between the biological, psychological and social factors, which in turn motivated me to engage passionately in the study and value the importance of addressing any group of people whose voice had not yet been heard within this academic context.

#### 4.4.2. Reflexivity In Recruitment

In response to hearing parents' apprehensions about engaging in the research, I was conscious of not wanting to burden the potential participants. I found it challenging to respond to people's concerns without feeling as if I was 'selling' my research and potentially persuading them to engage in a process with which they did not feel comfortable. I have chosen not to report the reasons people gave when opting not to participate, as I felt this to be a breach of confidentiality. Reporting their reasons inadvertently includes them in the research without their consent. This dilemma led me to question who the research was for. I was keen to keep the parents and their needs at the forefront of my mind, and not become too preoccupied about anticipated judgement of my sample size.

#### 4.4.3. Reflexivity In Interviewing

As discussed previously, I was aware that assumptions about my role within the London specialist paediatric hospital may have hindered or strengthened people's desire to discuss their experiences of accessing formal support within the health setting. This consideration led me to reflect further on the assumptions they may have held of me personally. I noticed I was asked several times if I had children. I decided to be transparent and disclose that I did not, as I wanted to promote a reciprocal relationship of trust. I did not explore this further,

but wondered whether, if I had children, parents might have felt more validated because I could better understand their devastation. Conversely, could my not being a parent allow them to be truly focused on their own devastation without being concerned regarding my potential experience of child health difficulties.

#### 4.4.4. Reflexivity In Analysis

Whilst thematic analysis does not explicitly prompt the researcher to consider their role as an active participant within the research process, I valued the opportunity to do so. As well as understanding that reflexivity is seen to promote rigorous and valid qualitative research (Harper, 2012), my position as a trainee Clinical Psychologist also underpinned the importance I placed on being reflexive whilst engaging in analysis.

Drawing from my experiences of designing the study, I was aware that my previous interactions with parents from different academic and clinical contexts might dominate my analysis and the expectations I had about the findings. To help ensure this did not interfere with my analysis, I drew from my clinical therapeutic experience which prompted me to stay curious throughout and allow for novel information to arise. Interestingly, I had not expected how challenging the parents' experiences of educational settings were, despite this having been a dominant theme within my previous research. I wondered whether my professional move from education to health influenced my frame of reference. I was surprised at how easily I had forgotten the complexity of educational systems. This challenge may exemplify the potential difficulty other professionals might have in holding the entirety of parents' experiences and how easy it can be to dismiss the significance of experiences in which you are not directly involved.

Through the multiple readings and coding of the transcripts, I developed a genuine admiration of the participants and their experiences. I became more emotionally connected to the content as the analysis progressed, perhaps as a result of becoming pregnant after completing the interviews. This helped me appreciate the experience of devastation the parents described, as the desire to have a healthy child is powerful. However, I was curious to notice my own expectations about my child's future were not influenced by the findings of the

current work or by my role as a trainee Clinical Psychologist within a paediatric oncology service.

#### 4.4.5. Reflexivity In Reporting and Dissemination

I initially found it challenging to move away from a descriptive account, and to develop themes. At times I felt uncomfortable collapsing the codes as I was worried I was losing the authenticity and 'voice' of the participant. To counter this, I attempted to use their words as accurately as I could. I kept the participants in mind throughout my writing, imagining what they might think of my treatment of their stories. Research and peer supervision also helped, as discussion and peer-reading ensured that the core of the participants' voices could still be heard.

At the time of writing this report I have already shared preliminary findings with the clinical multidisciplinary team who supported the study. I found their feedback interesting, with many clinicians expressing expectations the findings. One expectation was that the emotion of 'hope' would be more significant because recovery is possible. Although my findings did not suggest that parents never experience this, it felt important to communicate that, for most of the parents interviewed, the behavioural challenges and deterioration in health presentation dominated, leaving little space to consider the future and possible recovery.

One medical professional commented on the complexity of the parents' experiences compared with parents of children with cancer, "who go through predictable stages". This perspective underlined the simplistic framework that professionals outside the psychology context may use. Further feedback suggested that by highlighting some participants' perceived guilt about having financial resources that promoted better health outcomes for their child, I may have been presenting 'a bias against wealthy people'. I was at first confused, but through discussions with my academic supervisor I began to understand that wider medical discourse may find it uncomfortable to consider how social factors can impact health outcomes. In turn this may stem from an overarching discomfort in acknowledging societal and economic privilege (Afuape, 2012, p. 112).

#### **4.5. Conclusion**

Through the adoption of a qualitative design, parents shared a wide range of coping experiences associated with having a child diagnosed with LKS. The findings address the current gap within the literature and present a detailed account of the specific challenges faced across the course of LKS and the factors that helped parents cope. My research questions focused on the parental experience, and the outcomes highlighted several recurring themes including loss, powerlessness, knowledge, support, and strategies to cope.

The parental challenges experienced emphasised the significance of loss, with parents grieving the child once expected. This loss was further exacerbated by the rarity of the disorder, as lack of LKS knowledge made accessing helpful formal and informal support challenging. The feeling of powerlessness was dominant throughout parents' earlier experiences, as they watched their child's health deteriorate with no clear expectations about treatment and future health outcomes.

Reports of the consequential changes to the parents' lives across the course of the disorder was vast, with implications for parenting style, finances and social interaction. The need to take an advocate stance as a parent was also a dominant feature of parental reports, expressing the importance and complexity of developing a sense of power, control and authority.

Consequently, valuable support was understood by parents as gaining access to relevant knowledge, underpinned by experiences of not being judged and having their experiences validated. Therefore, the interaction with others did not necessarily need to be 'positive' in content, but needed to reflect an authenticity that led the parent to feel heard in some way.

The range of ways parents coped emphasised the importance of taking control. Whether taking charge of the role they played, their concept of time or the wider explanations for why LKS came in to their lives, the control gained through these

strategies exemplified the desire to create meaning within their own context and fight against the position of powerless expressed from the initial onset of LKS.

Drawing from key psychological factors and models, the findings were understood in the context of micro-to macro-level influences. The influence of wider cultural and societal contexts establishes norms which define 'typical' child development and parental responsibility. These influences parents' expectations in relation to parenting their child and the support desired. Additionally, these norms create expectations that lead to internal and external reactions, which can result in a complex interplay of emotions. Exemplified within the current study, parents' experiences encompassed descriptions of love, frustration, devastation and guilt.

In conclusion, this study will be beneficial to future parents and professionals who encounter LKS, as it presents the complexities of the parental experience and the unique interplay between the biological, psychological and social factors of LKS.

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## 6. APPENDICES

### Appendix A. Literature Search

The following search terms were used to access the literature surrounding families' experiences of LKS and wider parental experiences of other childhood health disorders that had relevant, overlapping clinical presentations (rarity of the disorder; the regression or loss of previously acquired cognitive functions; and behaviour which can be experienced as challenging). Literature was identified within the databases EBSCOhost, PsychARTICLES, PsychINFO and Science Direct. All article titles were scanned for relevance, and on this basis the relevant articles were read.

#### 1) ESBSCO PsychINFO and PsychARTICLES.

Date parameters: 1980 to 2017

Search Term 1	Search Term 2	Number of Articles
Landau Kleffner Syndrome	-	209
Landau Kleffner Syndrome	Experiences	8
Landau Kleffner Syndrome	Parents	8
Landau Kleffner Syndrome	Parents Experiences	1
Landau Kleffner Syndrome	Challenges	6
Landau Kleffner Syndrome	Coping	0
Rare disorders	Parents Experiences	9
Rare paediatric disorders	Parents Experiences	15
Rare paediatric epilepsy	Parents Experiences	2
Childhood Acquired Brain Injury	Parents Experiences	4
Paediatric Brain Injury	Parents Experiences	2
Autism Spectrum Disorder	Parents Experiences	339
Autism Spectrum Disorder	Parental challenges	8
Autism Spectrum Disorder	Parental coping	40

2) Science Direct

Date parameters: 1980 to 2017

<b>Search Term 1</b>	<b>Search Term 2</b>	<b>Number of Articles</b>
Landau Kleffner Syndrome	-	1,436
Landau Kleffner Syndrome	Experiences	0
Landau Kleffner Syndrome	Parents	600
Landau Kleffner Syndrome	Parents Experiences	382
Landau Kleffner Syndrome	Challenges	370
Landau Kleffner Syndrome	Coping	62
Rare disorders	Parents Experiences	32,564
Rare childhood disorders	Parents Experiences	18,013
Rare childhood disorders	Parental challenges	<b>5,016</b>
Rare childhood disorders	Parental coping	<b>2,477</b>
Rare paediatric disorders	Parents Experiences	<b>13,653</b>
Rare paediatric disorders	Parental challenges	<b>3,766</b>
Rare paediatric disorders	Parental coping	<b>1,428</b>
Rare paediatric epilepsy	Parents Experiences	<b>2,996</b>
Rare paediatric epilepsy	Parental challenges	<b>812</b>
Rare paediatric epilepsy	Parental coping	<b>320</b>
Childhood Acquired Brain Injury	Parents Experiences	<b>5,318</b>
Childhood Acquired Brain Injury	Parental challenges	<b>1,770</b>
Childhood Acquired Brain Injury	Parental coping	<b>972</b>
Autism Spectrum Disorder	Parents Experiences	<b>6,920</b>
Autism Spectrum Disorder	Parental challenges	<b>2,551</b>
Autism Spectrum Disorder	Parental coping	<b>1,136</b>

A snowballing effect from relevant articles was utilised, looking for appropriate literature on their reference lists. Additionally, Google Scholar and grey literature were also examined for relevant documents utilising the above search terms.



**Address**

**University of East  
London**  
School of Psychology  
Stratford Campus  
Water Lane  
London E15 4LZ

**Great Ormond Street Hospital**  
Great Ormond Street  
London  
WC1N 3JH

DATE

Dear

I am writing to request your participation in a study exploring parents' experiences of having a child diagnosed with Landau Kleffner Syndrome.

This research is being conducted in conjunction with the Developmental Epilepsy Clinic within Great Ormond Street Hospital and the University of East London.

Following discussions with Dr Maria Clark (Consultant Paediatric Neurologist), I would greatly appreciate the opportunity to interview you on your experiences. This research aims to further develop our understanding of Landau Kleffner Syndrome and inform how services and professionals can best support families.

If you would be interested in participating, please find a detailed information sheet enclosed. To arrange an interview or to ask any further questions about the study, please contact myself or my colleague named below.

If I do not hear from you in the next two weeks, I will contact you via telephone. This is to ensure that you received the letter and give further opportunity to discuss any questions or concerns you may have about participating.

Yours Sincerely,

Cleo Williamson  
Trainee Clinical Psychologist  
University of East London  
Email: u1438334@uel.ac.uk

Dr (Clinical Supervisor)  
Higher Specialist Clinical Psychologist  
Great Ormond Street Hospital

Appendix C. Ethical approval from Great Ormond Street Hospital's (GOSH) Clinical Research Adoptions Committee.

Please note that it is a GOSH formality that approval letters are addressed to the internal clinical lead, although I made the application.



Joint Research and Development Office  
Division of Research and Innovation

23.05.2016

Dr Rebecca Greenaway  
Higher Specialist Clinical Psychologist  
Great Ormond Street Hospital

Dear Dr Greenaway

PI: Dr Rebecca Greenaway  
R&D Number: 16HN09  
Title: Parents' Experiences of having a child diagnosed with Landau Kleffner Syndrome  
Funding: There is no funding allocated to this project

I am writing to inform you that the Clinical Research Adoptions Committee (CRAC) reviewed your application and has no objections to the conduct of this project at GOSH although you will need to obtain ethics approval. Overall this looks a good application. The Committee had a couple of comments and suggestions:

- The application refers to the importance of early identification and early intervention. It might therefore be helpful and informative to include questions about when parents first noticed differences, what this looked like and what their experiences were regarding accessing early intervention.
- The committee recommends considering increasing the numbers slightly. Guest, Bunce and Johnson (2006) are quoted as suggesting a sample of 12 will be sufficient for such studies. The stated intention is to interview 12-20 parents (which could relate to experiences regarding 6-10 patients). Given parents/carers of any one child may share experiences to some extent, aiming for 20+ might be more appropriate to ensure richness of themes generated.
- The information sheet should read parents and carers and not mother and fathers.

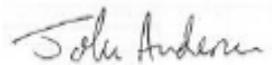
Joint Research and Development Office  
Division of Research and Innovation  
UCL Institute of Child Health, 30 Guilford Street, London WC1N 1EH  
Tel: 020 7906 2700 Fax: 020 7906 2201  
www.gosh.nhs.uk

The child first and always

**Decision: Approval**

You will shortly be contacted by R&D Governance who will support you through the process of obtaining the necessary approvals before your project can begin. You must not commence your project before receiving R&D approval. Please find attached further information regarding the next stages in the research administration process.

Regards,



**Chair**  
**Clinical Research Adoption Committee**

## Appendix D. Ethical approval from the Westminster Research Ethics Committee



### Health Research Authority

London - Westminster Research Ethics Committee  
4 Minshull Street  
Manchester  
M1 3DZ

Telephone: 0207 104 8012

**Please note: This is the favourable opinion of the REC only and does not allow you to start your study at NHS sites in England until you receive HRA Approval**

30 September 2016

Miss Cleo Williamson  
50 Southstand Highbury Stadium Square  
London  
N5 1EY

Dear Miss Williamson

**Study title:** PARENTS' EXPERIENCES OF HAVING A CHILD  
DIAGNOSED WITH LANDAU KLEFFNER SYNDROME  
**REC reference:** 16/LO/1484  
**IRAS project ID:** 210547

Thank you for your submission of 13 September 2016, responding to the Committee's request for further information on the above research and submitting revised documentation.

The further information has been considered on behalf of the Committee by the Alternate Vice-Chair, Dr Erika Kennington and Miss Rachel Fay.

We plan to publish your research summary wording for the above study on the HRA website, together with your contact details. Publication will be no earlier than three months from the date of this opinion letter. Should you wish to provide a substitute contact point, require further information, or wish to make a request to postpone publication, please contact the REC Manager, Ms Rachel Katzenellenbogen, [nrescommittee.london-westminster@nhs.net](mailto:nrescommittee.london-westminster@nhs.net).

#### Confirmation of ethical opinion

On behalf of the Committee, I am pleased to confirm a favourable ethical opinion for the above research on the basis described in the application form, protocol and supporting documentation as revised, subject to the conditions specified below.

### **Conditions of the favourable opinion**

The REC favourable opinion is subject to the following conditions being met prior to the start of the study.

Management permission must be obtained from each host organisation prior to the start of the study at the site concerned.

Management permission should be sought from all NHS organisations involved in the study in accordance with NHS research governance arrangements. Each NHS organisation must confirm through the signing of agreements and/or other documents that it has given permission for the research to proceed (except where explicitly specified otherwise).

Guidance on applying for NHS permission for research is available in the Integrated Research Application System, [www.hra.nhs.uk](http://www.hra.nhs.uk) or at <http://www.rforum.nhs.uk>.

Where a NHS organisation's role in the study is limited to identifying and referring potential participants to research sites ("participant identification centre"), guidance should be sought from the R&D office on the information it requires to give permission for this activity.

For non-NHS sites, site management permission should be obtained in accordance with the procedures of the relevant host organisation.

Sponsors are not required to notify the Committee of management permissions from host organisations.

### Registration of Clinical Trials

All clinical trials (defined as the first four categories on the IRAS filter page) must be registered on a publically accessible database within 8 weeks of recruitment of the first participant (for medical device studies, within the timeline determined by the current registration and publication trees).

There is no requirement to separately notify the REC but you should do so at the earliest opportunity e.g. when submitting an amendment. We will audit the registration details as part of the annual progress reporting process.

To ensure transparency in research, we strongly recommend that all research is registered but for non-clinical trials this is not currently mandatory.

If a sponsor wishes to contest the need for registration they should contact Catherine Blewett ([catherineblewett@nhs.net](mailto:catherineblewett@nhs.net)), the HRA does not, however, expect exceptions to be made. Guidance on where to register is provided within IRAS.

**It is the responsibility of the sponsor to ensure that all the conditions are complied with before the start of the study or its initiation at a particular site (as applicable).**

### **Ethical review of research sites**

#### NHS sites

The favourable opinion applies to all NHS sites taking part in the study, subject to management permission being obtained from the NHS/HSC R&D office prior to the start of the study (see "Conditions of the favourable opinion" below).

#### Approved documents

The final list of documents reviewed and approved by the Committee is as follows:

Document	Version	Date
Evidence of Sponsor insurance or indemnity (non NHS Sponsors only) [UEL Certificate]	Version 2	11 September 2016
Interview schedules or topic guides for participants [Interview Schedule]	1	11 September 2016
IRAS Application Form [IRAS_Form_21072016]		21 July 2016
Letters of invitation to participant [Invite to participants]	2	11 September 2016
Other [Debrief Sheet]	1	19 June 2016
Other [Professional Indemnity Policy Schedule]		11 September 2016
Participant consent form [Consent form]	1	19 June 2016
Participant consent form [Consent form]	2	13 September 2016
Participant information sheet (PIS) [Information Sheet]	2	11 September 2016
Referee's report or other scientific critique report [GOSH CRAC decision]		23 May 2016
Research protocol or project proposal	1	19 June 2016
Summary CV for Chief Investigator (CI)		
Summary CV for supervisor (student research) [Academic S CV]		24 June 2016

#### Statement of compliance

The Committee is constituted in accordance with the Governance Arrangements for Research Ethics Committees and complies fully with the Standard Operating Procedures for Research Ethics Committees in the UK.

#### After ethical review

##### Reporting requirements

The attached document "*After ethical review – guidance for researchers*" gives detailed guidance on reporting requirements for studies with a favourable opinion, including:

- Notifying substantial amendments
- Adding new sites and investigators
- Notification of serious breaches of the protocol
- Progress and safety reports
- Notifying the end of the study

The HRA website also provides guidance on these topics, which is updated in the light of changes in reporting requirements or procedures.

#### **User Feedback**

The Health Research Authority is continually striving to provide a high quality service to all applicants and sponsors. You are invited to give your view of the service you have received and the application procedure. If you wish to make your views known please use the feedback form available on the HRA website:

<http://www.hra.nhs.uk/about-the-hra/governance/quality-assurance/>

#### **HRA Training**

We are pleased to welcome researchers and R&D staff at our training days – see details at

<http://www.hra.nhs.uk/hra-training/>

**16/LO/1484**

**Please quote this number on all correspondence**

With the Committee's best wishes for the success of this project.

Yours sincerely



**Mr Robert Goldstein**  
Acting Chair

Email: [nrescommittee.london-westminster@nhs.net](mailto:nrescommittee.london-westminster@nhs.net)

Enclosures: "After ethical review – guidance for researchers"

Copy to: Professor Neville Punchard, University Of East London  
Dr Thomas Lewis, Great Ormond Street Hospital for Children NHS  
Foundation Trust

## Appendix E. Ethical approval from the Health Research Authority



Health Research Authority

Miss Cleo Williamson  
50 Southstand Highbury Stadium Square  
London  
N5 1EY

Email: [hra.approval@nhs.net](mailto:hra.approval@nhs.net)

28 October 2016

Dear Miss Williamson

### Letter of HRA Approval

**Study title:** PARENTS' EXPERIENCES OF HAVING A CHILD DIAGNOSED WITH LANDAU KLEFFNER SYNDROME  
**IRAS project ID:** 210547  
**REC reference:** 16/LO/1484  
**Sponsor** University Of East London

I am pleased to confirm that HRA Approval has been given for the above referenced study, on the basis described in the application form, protocol, supporting documentation and any clarifications noted in this letter.

#### Participation of NHS Organisations in England

The sponsor should now provide a copy of this letter to all participating NHS organisations in England.

*Appendix B* provides important information for sponsors and participating NHS organisations in England for arranging and confirming capacity and capability. Please read *Appendix B* carefully, in particular the following sections:

- *Participating NHS organisations in England* – this clarifies the types of participating organisations in the study and whether or not all organisations will be undertaking the same activities
- *Confirmation of capacity and capability* - this confirms whether or not each type of participating NHS organisation in England is expected to give formal confirmation of capacity and capability. Where formal confirmation is not expected, the section also provides details on the time limit given to participating organisations to opt out of the study, or request additional time, before their participation is assumed.
- *Allocation of responsibilities and rights are agreed and documented (4.1 of HRA assessment criteria)* - this provides detail on the form of agreement to be used in the study to confirm capacity and capability, where applicable.

Further information on funding, HR processes, and compliance with HRA criteria and standards is also provided.

It is critical that you involve both the research management function (e.g. R&D office) supporting each organisation and the local research team (where there is one) in setting up your study. Contact details

IRAS project ID	210547
-----------------	--------

#### HRA Training

We are pleased to welcome researchers and research management staff at our training days – see details at <http://www.hra.nhs.uk/hra-training/>

Your IRAS project ID is **210547**. Please quote this on all correspondence.

Yours sincerely

Alison Thorpe  
Senior Assessor

Email: [hra.approval@nhs.net](mailto:hra.approval@nhs.net)

*Copy to: Professor Neville Punchard, University Of East London  
Dr Thomas Lewis, Great Ormond Street Hospital for Children NHS Foundation  
Trust*



## Participant Information Sheet

The purpose of this letter is to provide you with the information that you need to consider in deciding whether to participate a research study. The study is being conducted as part of my Professional Doctorate in Clinical Psychology degree at the University of East London.

**Project Title:** Parents' Experiences of Having a Child Diagnosed with Landau Kleffner Syndrome

### **What Is the Purpose Of The Study?**

Most research on Landau Kleffner Syndrome (LKS) focuses on biological descriptions, with little attention given to the affect this condition may have on the family. I will be interviewing mothers and fathers whose children were given a diagnosis of LKS in childhood, and asking about the possible stressors they have faced and the factors that may have helped them to cope. The project hopes to further develop our understanding of LKS and inform how services and professionals can best support families. The study is being conducted as part of my Professional Doctorate in Clinical Psychology at the University of East London.

### **What Would Taking Part Involve?**

I shall visit you at a place of your choosing to conduct an interview. Ideally the interviews need to be conducted; individually, face to face, in a quiet environment, with minimal distractions. Unfortunately, no costs towards child care can be provided.

I am based in London, so the distance to your preferred location may need to be taken into account when arranging a suitable time. With your consent, the interview will be recorded and transcribed.

### **What are the possible benefits of taking part?**

This research will give you the opportunity to talk about your experiences, contribute to potential future service development and promote wider awareness of the disorder.

### **What are any possible disadvantages and risks of taking part?**

The study is not anticipated to cause anyone distress however it may be upsetting talking about experiences that were found to be particularly difficult. For this reason, interviews can be paused or stopped at any time, and access to support can be discussed.

### **Will my taking part in this study be kept confidential?**

All interviews will be recorded and transcribed into word documents. These recordings will be password protected, whilst the transcripts will be anonymised with pseudonyms. Interview transcripts will only be available to the researcher, supervisor and thesis examiner. Electronic data will be stored on a desktop

computer at Great Ormond Street Hospital, where all physical and IT security measures are provided. Any paper or manual files will be stored in a locked cabinet, with access only available for clinicians involved in the research. In accordance with the Data Protection Act (1998), all data will be deleted after 5 years.

The only instance where confidentiality could be broken would be if I had concerns about risk involving yourself or another person. In this instance I would inform you and my supervisor of my concerns, and a plan of action would be discussed.

**What will happen to the results of the research study?**

Before the data is analysed, you will have the opportunity to review your interview transcript and make corrections. The results of the research study will be written up and form the basis of my Professional Doctorate thesis. Parts of the study will also be submitted for publication. Both the thesis and publication, will include direct quotes from the interviews.

**What will happen if I don't want to carry on with the study?**

You are not obliged to take part in this study and should not feel coerced. You are free to withdraw at any time. Should you choose to withdraw from the study you may do so without disadvantage to yourself and without any obligation to give a reason. Should you withdraw after the interview has been transcribed, the researcher reserves the right to use your anonymised data in the write-up of the study and any further analysis that may be conducted by the researcher.

Please feel free to ask me any questions. If you are happy to continue you will be asked to sign a consent form prior to your participation. Please retain this invitation letter for reference.

If you have any questions or concerns about how the study has been conducted, please contact the project supervisor Dr Emma McGibbon, Specialist Clinical Psychologist within the Neurodisability Service, Great Ormond Street Hospital, Great Ormond Street Hospital for Children NHS Foundation Trust, Great Ormond Street, London WC1N 3JH, Telephone. 020 7405 9200

**or**

Chair of the School of Psychology Research Ethics Sub-committee: Dr. Mary Spiller, School of Psychology, University of East London, Water Lane, London E15 4LZ.

(Tel: 020 8223 4004. Email: [m.j.spiller@uel.ac.uk](mailto:m.j.spiller@uel.ac.uk))

Thank you in anticipation.

Yours sincerely,

Cleo Williamson  
Principle investigator

Appendix G. Participant Consent Form



**CONSENT FORM**

Title of Project: Parents' Experiences Of Having A Child Diagnosed With Landau Kleffner Syndrome

Name of Researcher: Cleo Williamson

Please initial box

- 1. I confirm that I have read the information sheet dated..... (version.....) for the above study. I have had the opportunity to consider the information, ask questions and have had these answered satisfactorily.
- 2. I understand what is being proposed and the procedures in which I will be involved have been explained to me.
- 3. I understand that my involvement in this study, and particular data from this research, will remain strictly confidential.
- 4. It has been explained to me what will happen once the research study has been completed and that direct quotations from the interviews can be used in future publications.
- 5. I understand that my participation is voluntary and that I am free to withdraw at any time without giving any reason.
- 6. I also understand that should I withdraw after the interview has been transcribed, the researcher reserves the right to use my anonymous data in the write-up of the study and in any further analysis that may be conducted by the researcher.
- 7. I agree to take part in the above study.

\_\_\_\_\_  
Name of Participant                      Date                      Signature

\_\_\_\_\_  
Name of Person                      Date                      Signature  
taking consent



**UNIVERSITY OF EAST LONDON**

School of Psychology  
Stratford Campus  
Water Lane  
London E15 4LZ

Thank you for taking part in this study. Your time and effort are much appreciated. Within this study we looked at your experience of having a child with a diagnosis of Landau Kleffner Syndrome.

Whilst the study is for my doctorate thesis, it is hoped that this work will be published and assist services in developing ways to best support families whose children are given a diagnosis of Landau Kleffner Syndrome. You are welcome to request a final copy of the thesis by contacting me.

If you have experienced any distress following the interview or would like to further discuss issues raised, please contact myself or Dr Emma McGibbon on;

[U1438334@uel.ac.uk](mailto:U1438334@uel.ac.uk)

[Emma.McGibbon@gosh.nhs.uk](mailto:Emma.McGibbon@gosh.nhs.uk)

Yours sincerely,

Cleo Williamson

## Appendix I. Interview schedule

### **Interview Schedule**

#### Project Title: Parents' Experiences of Having a Child Diagnosed with Landau Kleffner Syndrome

The information sheet to be discussed, ensuring the projects aims are clear, and to answer any questions/queries they may have.

Confidentially and consent discussed, with consent form signed.

Collection of demographic details of participant and the child being discussed.

Questions aim to be in chronological order of experience. However, the interview is to be participant led and flexibility will be given.

- Can you describe the events that led to you discovering that your child had difficulties?
- Can you describe the events leading up to your child gaining a diagnosis LKS?
- How did you find this process?
- Has the 'management' of having a child with LKS changed over the years?
- Have particular ages/milestones been easier or harder than others?
- What reactions have you had from family and friends?
- Would you say you have a 'support system' of some kind, formal or informal?
- Would there be any advice or knowledge you would pass on to another parent?
- Would this advice conflict with your own actions?
- Do you have any specific thoughts about the future?

Thank the individual for participating

Appendix J. Transcript Convention Adapted from Parker (2005)

**P** Indicates participant

**I** Indicates interviewer

**( )** Indicates pause in speech

**[unclear]** Indicates speech was unclear

**[ ]** Indicates when a comment has been added by the author

**< >** Indicates interruption

**/** Indicates overlapping speech

**-** Indicates unfinished word

## Appendix K. Coded transcript

261	Math and things like that were just very very visual () keeping everything visual. And also I	<b>Author</b>
262	thought it's really important when you first got to hospital to tell them all the things he could	Acknowledge Positives
263	do still, not just the stuff he couldn't do. He could do a lot.	
264		<b>Author</b>
265	PB: And again I feel we were really lucky because his Landau Kleffner was very pure in	Lucky – acknowledging positives
266	terms of being just aphasia. Physically he was fine, he could still ride his bike. He could	<b>Author</b>
267	still go out and run around. Other children didn't want anything to do with him, but he could	Watching social exclusion
268	still do it.	<b>Author</b>
269		Childs desire to interact
270	PB: Socially and emotionally he still wanted to engage he didn't cut off or anything like	<b>Author</b>
271	that. Which really helped. I think when he was in high school. To have those friendships	Child can't remember
272	and things. Because I think. I mean he says he doesn't really remember an awful lot of it.	<b>Author</b>
273		Watching social isolation
274	PB: But one of the other hardest aspects is just to see a child who's naturally so sociable	<b>Author</b>
275	be so isolated and lonely. We had two lovely boys who lived next door and he would	Child desire to interact
276	kidnap them whenever he could. They did spend a lot of time here and he really didn't	<b>Author</b>
277	want them to go. I mean he was so desperately craving that contact. He had lots of laughs	His positive experiences - Positives
278	as well.	
279		
280	I: In those moments when he had his friends round, they got on well?	<b>Author</b>
281		Use of humor- focusing on the positives
282	PB: They really did, by this time he did have quite a lot of language and there were lots of	<b>Author</b>
283	errors in his speech and things, but they just used to fall about laughing. And so he would	Other's input helped him socially
284	fall out laughing which I found really helpful because I wouldn't naturally fall about	<b>Author</b>
285	laughing. I decipher and things like that. That really helped him. And they play games and	Importance of holding on to positives
286	stuff as well.	<b>Author</b>
287		Memory of the healthy child. LKS traps child
288	I: As you're talking is sound like you really value holding onto the positives of those	<b>Author</b>
289	moments.	Invisibility
290		<b>Author</b>
291	PB: Absolutely, definitely and really important I think it was. I mean I think it is the nature of	Lack of parental awareness
292	the illness. Because your child has actually been doing well and to a certain point you	<b>Author</b>
293	never lose sight of that. Healthy child. Well I didn't anyway. It really did feel like he was	Invisibility- lack of parental awareness
294	completely trapped by this horrible illness. And it was such a difficult illness to get your	<b>Author</b>
295	head round because he never had dropped down seizures who never had seizures that	Witnessing the deterioration
296	other people would see or I would see. So you literally had no idea.	<b>Author</b>
297		Lack of awareness or knowledge about presentation
298	PB: You know I was put into bed at night before the diagnosis I thought, oh he's going to	<b>Author</b>
299	have good night's sleep and maybe tomorrow he'll be better. But he wasn't having a good	Organizational challenges
300	night's sleep. He was having seizures eighty percent of the time he was asleep. And ... But	<b>Author</b>
301	I didn't know that. I didn't know why he started wetting the bed again. And that was a	Challenges of organizing treatment
302	horrible regression before diagnosis. And I would never have guessed that it was epilepsy	<b>Author</b>
303	because there isn't any in my family that I know.	Being proactive
304		<b>Author</b>
305	PB: Another really demanding side of it. was keeping on top of on a daily basis. I mean I	Organizational- full time job
306	have reams and reams of diaries. Everyday I would write down what med's he'd had, if	<b>Author</b>
307	he'd done anything quirky. What I needed to do to chase it up. When the meds had to go	Child awareness- acknowledgment
308	up, when the meds had to go down. And if I hadn't of written it down I don't know how -.	<b>Author</b>
309	Organisationally it was Demanding, it was like full time job. Got two huge box files of	Holding primary responsibility
310	correspondents with schools trying to find schools. And I've kept them all. Because I	
311	thought one day X may want to trawl through all that. But he might want to see some of it.	
312	Organisationally it was hard work. And that was down to me.	

## Appendix L. Initial Codes and Frequencies

The table below displays the initial codes developed from annotating the raw data transcripts. The sources column indicates the number of participants that mentioned each code.

<b>Code no.</b>	<b>Initial Codes</b>	<b>Source</b>
1.	Previous typical ability	8
2.	Typical development	5
3.	Change in behaviour	6
4.	Dangerous behaviour	5
5.	Change in health	3
6.	Loss of skills	5
7.	Loss and acquisition of skills	5
8.	Invisible	2
9.	Child not remembering past	4
10.	Childs loss of confidence	2
11.	Child being socially excluded	2
12.	Shy with other children	2
13.	Aggressive with other children	3
14.	Current difficulties in cognitive abilities	3
15.	Childs desire to interact	3
16.	Parental distress	2
17.	Parental fear	6
18.	Parental upset	3
19.	Lack of knowledge about presentation	4
20.	Attempt to find cause	4
21.	Teacher becoming initially aware of difficulty	1
22.	Not findings initially found	3
23.	Initial diagnosis of epilepsy	4
24.	Initial misdiagnosis	3
25.	Concerns about the future	3
26.	Confusion about diagnosis	3
27.	Unhelpful to receive diagnosis	2
28.	Medication given	2
29.	Negative impact of medication treatment	6
30.	Positive experiences of medication treatment	2
31.	Operation positives	1
32.	Operations negatives	1
33.	Advice to be assertive	4
34.	Example of being assertive	4
35.	Hard being assertive	2
36.	Observing other parents doing less	6

37.	Child needs me to advocate	3
38.	Worried health would get worse	2
39.	Memories of previous presentation	2
40.	No boundaries	1
41.	No behaviour problems	1
42.	Not coping	3
43.	Protective	4
44.	Restraint	2
45.	Structure	2
46.	Worry about siblings	2
47.	Impact on siblings	1
48.	Seeking support for behaviour	3
49.	Difficulty making plans	3
50.	Changes to employment	2
51.	Financial strain	4
52.	Impact on wider family	4
53.	Impact on spousal relationship	2
54.	Organisation	2
55.	Time feeling longer	1
56.	Acknowledging positives	6
57.	Positive treatment outcomes	5
58.	Others have it worse	3
59.	Having financial resources	3
60.	Being proactive	4
61.	Got stronger	2
62.	Religious explanation	2
63.	One day at a time	2
64.	Memory fading	3
65.	Hard to get an appropriate school	3
66.	Hard to get a statement of educational needs	3
67.	School having a lack of knowledge	2
68.	School not following guidance	2
69.	School -poor management of child's behaviour	2
70.	Good academic development	3
71.	Helpful home support	1
72.	Helpful behaviour management	1
73.	Child's social skills improving	2
74.	School understanding child's needs	3
75.	Judgement from professionals about behaviour	3
76.	Unhelpful behaviour strategies given	2
77.	Helpful behaviour strategies	3
78.	Unhelpful speaking to other parents (LKS)	3

79.	Helpful speaking to other parents (LKS)	4
80.	Grandparents helpful	4
81.	Parent holding main responsibility	5
82.	Spouse being a support	3
83.	Differences in spouses understanding	4
84.	Judgment from others	5
85.	Positive social support	3

## Appendix M. Examples of Coded Data Extracts

<p>6. Loss of Skills</p>	<p>P1: he couldn't count anymore, couldn't use a knife and fork, wetting himself, before everything was fine. Then Little things they pick up, singing, things like singing along to nursery rhymes or things that you know, they all change. He literally changed. As a child. He went from absolutely angelic, slept, ate, I couldn't have been luckier as a first time mum if I'd tried. Everything was perfect, great fun child no anger issues no temper tantrums nothing. and then progressively, went 10 to 0 over over 18 months. and practical zero output as well. So he was a mess really.</p> <p>P3: She'd forgets things, like if the phone would ring she wouldn't pick up whereas before she would run to the phone.</p> <p>P4: I went on holiday with the girlfriends in July. And when I came back, I was gone for two weeks, his language had completely changed. Because I'd been away from him, and came back, I said to husband , he's not pronouncing his words properly. He's slurring his words. And within about a month he couldn't talk.</p> <p>P5: Well I think he was poorly or were we were worried about it. because he wasn't hearing or speaking</p> <p>P8: Around that time we first got to the hospital got the diagnosis of LKS and he had zero verbal comprehension</p>
<p>17. Parental Fear</p>	<p>P1: So I picked him up and he was obviously ridged, rock solid. And connected to the mobile home park there is a pub and restaurant. So we ran straight up there. Just panicking because we knew a nurse was there. We honestly thought he was dying.</p> <p>P2: I did ask him [doctor], what am I looking at. And he said what are you asking me? I was like what what's going? And he said are you asking me if she's going to die? I don't know am I. And he said if you didn't get her here when you did, she probably would. (became upset)</p> <p>P4: I'd be calling the ambulance in the night because he couldn't breathe. He'd go blue. It was every single night. We'd have to take it in turns to see him because we were worried he might die. It was that bad.</p> <p>P5: ...I realized his bed was wet and I thought that its really unusual. That's a bit strange and changed his changed his bed, went to look at him and of course he was seizing and was quite blue. And I actually called an ambulance as I was quite concerned about him being so blue.</p> <p>P7: There was that worry that she would set fire to something because- it was always worrying, always worrying. You couldn't leave her for a second. I couldn't leave her to go to the toilet.</p> <p>P8: And I have a great GP so she asked what was my biggest fear. My biggest fear was he actually had a brain tumour or</p>

	something because it was clearly getting worse.”
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## Appendix N. Grouped Final Codes

