

# From Symptom Onset to Diagnosis:

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a critical exploration into the experiences of  
young people with Juvenile-onset Systemic  
Lupus Erythematosus

By

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A thesis submitted in partial fulfilment for the requirements for  
the degree of MSc by Research at the University of Central  
Lancashire.

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## **Student Declaration**

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I declare that while registered as a candidate for the research degree, I have not been a registered candidate or enrolled student for another award of the university or other academic or professional institution.

I declare that no material contained in the thesis has been used in any other submission for an academic award and is solely my own work.

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Type of Award .....  
School .....

## Abstract

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Juvenile Systemic Lupus Erythematosus (JSLE) is a life-long condition affecting 6-30 per 100,000 children and young people per year, depending on ethnicity, in the UK (Papadimitraki & Isenberg 2009). Symptoms include fatigue, tiredness, malar rash, joint problems, headaches and kidney problems. There is a paucity of literature relating to the experiences of young people before and during diagnosis of JSLE. An understanding of young people's experiences and the issues relevant to them could help improve outcomes and facilitate the development of standards of care in JSLE.

The aims of the study were to: (1) describe the journey from onset of symptoms to diagnosis in JSLE from a young person's perspective by exploring the stories they tell; and (2) ascertain key points, if any, in the journey to diagnosis to generate deeper insight into access to care for young people with JSLE.

This practitioner-researcher study used an exploratory qualitative approach. The setting was the UK tertiary centre for paediatric rheumatology in which the researcher works. The study utilised audio-recorded, in-depth interviews with eight young people (aged 12-19 years) with JSLE who told their story of their own 'journey to diagnosis'. Parental consent was obtained for their child's participation for young people aged under 16 years and assent was gained from every young person under the age of 16. Those young people aged 16 and above provided their own consent in accordance with ethical guidelines. The audio-recordings were transcribed and the transcripts were subjected to thematic analysis. The researcher maintained a fieldwork diary and used a reflexive approach throughout the study.

Four main themes were generated and these themes are linked by a meta-theme 'passing of time'. Although evident across all the themes, the 'passing of time' was not a static concept as it evolves and changes and influences the young people's experiences. The themes are chronologically ordered reflecting the young people's 'journey to diagnosis'. The first theme, 'Emerging Illness' encompasses the first descriptions of a change in health, the emergence of physical symptoms and the impact of these symptoms on the young people's lives. Often symptoms were dismissed or ignored. The second theme, 'Seeking Help' lasted from a period of several weeks to up to 2 years and covered the first and ongoing contacts with health services. Most young people experienced dissatisfaction with how their symptoms were dealt with. The third theme, 'Diagnosis of Lupus' was a significant time point as the young people experienced a major change in health status. For some, diagnosis was a relief; others were worried by the implications of the condition. Finally, the fourth theme, 'Resilience, Reflection & Recovery' encompasses the experiences that had occurred after diagnosis. It is characterised by things that went well and things that did not.

In conclusion, all young people should have the opportunity to tell their 'journey to diagnosis' story to a care team member. Understanding young people's experiences of 'symptom onset to diagnosis' has the potential to reduce the impact and burden of this disease. Key recommendations include supporting and educating young people to recognise symptom-related changes; improving mechanisms, especially within primary care, for young people and families seeking help for such symptoms; raising awareness of emerging symptoms of lupus in young people within the wider community, including schools; providing young people with the opportunity to reflect and tell their story from 'symptom onset to diagnosis' to an appropriate member of the healthcare team; increasing support, especially at school, for young people after they have been diagnosed to enable them to successfully negotiate this life long illness through adolescent transition into adulthood.

## Table of Contents

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### Contents

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Student Declaration.....	1
Abstract .....	2
Table of Contents .....	3
List of Tables and Illustrative Material .....	5
Acknowledgments .....	6
CHAPTER I: INTRODUCTION .....	7
CHAPTER II: LITERATURE REVIEW.....	10
Introduction .....	10
Problem Identification.....	11
Literature Search .....	12
Data Evaluation.....	14
Data Analysis .....	17
Presentation of Results .....	27
Conclusions.....	28
CHAPTER III: METHODOLOGY.....	30
Introduction.....	30
Aims of the study.....	30
Target Population .....	31
Recruitment and Consent.....	32
Methods .....	33
Data Analysis .....	36
Evaluation Using CASP .....	38
Summary.....	39
CHAPTER IV: FINDINGS.....	41
Introduction.....	41
Overview of key themes.....	43
Emerging Illness.....	43
Seeking Help .....	43
Diagnosis of Lupus.....	43
Resilience, Reflection and Recovery.....	44
Section One - Emerging Illness .....	45

Introduction.....	45
Emerging Physical Symptoms.....	45
Emerging Disruption to Life.....	48
Section Two – Seeking Help.....	51
Introduction.....	51
Experiences of Dissatisfaction.....	51
Feelings of Uncertainty.....	53
Section Three - Impact of Diagnosis of Lupus.....	55
Introduction.....	55
Impact on self.....	55
Impact on family.....	59
Section Four - Resilience, Reflection and Recovery.....	62
Introduction.....	62
Resilience.....	62
Reflection.....	63
Recovery.....	65
Conclusion.....	66
CHAPTER V: DISCUSSION.....	67
Introduction.....	67
Emerging Illness.....	68
Seeking Help.....	72
Diagnosis of Lupus.....	74
Resilience, Reflection & Recovery.....	77
Passing of Time.....	79
Limitations.....	80
Recommendations for Practice.....	82
Recommendations for Research.....	83
Conclusion.....	84
REFERENCES.....	85
APPENDICES.....	94
Appendix 1.....	95
Appendix 2.....	96
Appendix 3.....	97
Appendix 4.....	98
Appendix 5.....	99

## List of Tables and Illustrative Material

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- Table I. Search Strategy.....11
  - Table II. The data evaluation of the reviewed papers .....14-16
  - Table III. Classification Criteria for Systemic Lupus Erythematosus .....18
  - Table IV. Evaluation of the Study using CASP.....38-39
  - Table V. Demographic Information & Code Identifiers of Young People.....42
- 
- Figure 1: The Role of the UK JSLE Study Group .....8
  - Figure 2: The literature search process..... 13
  - Figure 3: Showing summary points of the synthesis of theoretical and empirical evidence and the implications for future research..... 28
  - Figure 4: The data analysis process.....37
  - Figure 5: Illustrating four main themes & their relationship to meta-theme.....68

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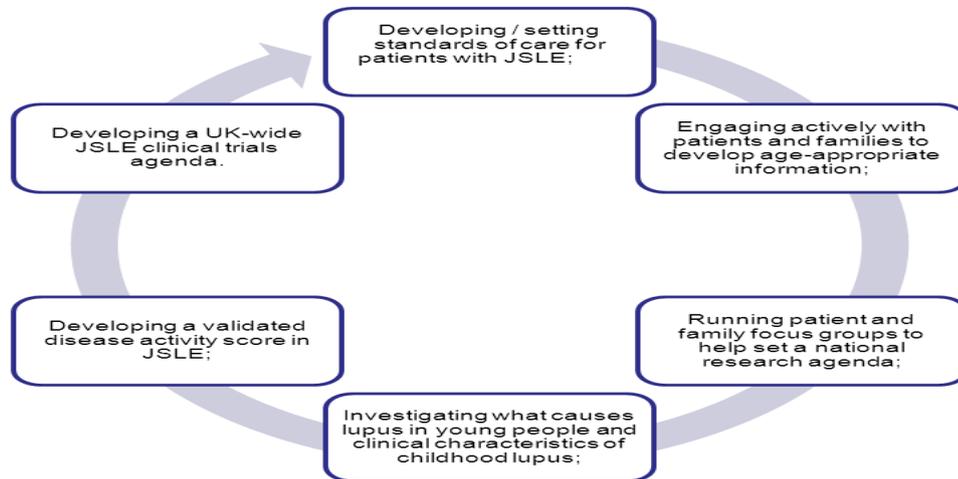
## CHAPTER I: INTRODUCTION

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Young people diagnosed with juvenile onset systemic lupus erythematosus (JSLE) a rare, severe, relapsing condition have to cope with the impact of significant illness at a time when they are growing and developing both physically and psychologically (Kelly et al. 2012). Medicines used to manage JSLE have notable side effects and co-morbidities frequently occur (Smith et al. 2012). There is a greater risk of severe organ damage in childhood lupus, in particular affecting the renal system compared to adult onset disease (Kamphuis & Silverman 2010; Watson et al. 2012). It is estimated that 20% of people with lupus are diagnosed during childhood (Tucker et al. 2008); approximately 6 to 30 per 100,000 children/ young people per year depending on ethnicity are affected in the United Kingdom (UK) (Papadimitraki & Isenberg 2009). Only approximately 500 children/ young people in the UK are affected. There is a higher ratio of females to males, around 5:1, diagnosed with JSLE compared to adult onset lupus where the ratio of females to males is much higher at approximately 9:1 (Tucker et al., 2008; Maidhof & Hilar, 2012; Watson et al. 2012). The period leading up to diagnosis can be particularly difficult for young people as they present with symptoms and seek a diagnosis that is both challenging and complex to make (Watson et al. 2011).

JSLE is an autoimmune multisystem disease that, although rare, can be aggressive and is associated with significant morbidity and has a higher risk of death than adult onset systemic lupus erythematosus (Watson et al 2012). Though there have been improvements in the understanding and management of JSLE the impact and burden of disease on children and young people diagnosed in childhood remains substantial. In 2006 the United Kingdom Juvenile Systemic Lupus Erythematosus Study Group was established to support a widespread programme of clinical research into JSLE (see Figure 1). Through the UK JSLE Cohort Study and Repository the group collects clinical data and blood samples from children across the UK who fulfil the SLE American College of Rheumatology (1997) classification for SLE<18 years of age and those with probable

evolving disease from time of diagnosis onwards. To date the cohort study has patients enrolled from 17 centres nationwide, 68 of whom have been recruited at a northwest tertiary centre (TC) for paediatric rheumatology. Ultimately, the focus of the UK JSLE Study Group is to improve the care of patients with JSLE.



**Figure 1: The Role of the UK JSLE Study Group** (Note: diagram adapted from UK JSLE Study Group poster, British Society of Paediatric and Adolescent Rheumatology Autumn Conference 2012).

Evidence from research into other childhood rheumatic conditions has revealed that timely access to specialist rheumatology services does not always occur and where delays are present disease outcomes can be significantly affected (Hawley et al. 2012, Foster and Rapley 2010). A referral to specialist rheumatology services has been described by families as critical in the process of diagnosis in Juvenile Idiopathic Arthritis (JIA) (Britton & Moore 2002). Further to this, families have reported that difficult ‘journeys to diagnosis’ have had a negative impact on the way they subsequently related to health care practitioners (Knafl et al. 1995). Thus, if the experiences of families are investigated, and the positives and negatives identified, this information could be used to plan a responsive specialist service that is aware of the needs of its families. The priorities of families who access health services have been shown to differ from those of professionals (Neill 2000). In JIA the period leading up to diagnosis was criticised by the majority of parents for its length and lack of speed between symptom onset and diagnosis (Barlow et al. 1998). The British Society of Paediatric and Adolescent Rheumatology (BSPAR)

have developed Standards of Care for Children and Young People with JIA (2010). An expert working group that included young people with arthritis and their parents met to determine evidence-based standards to meet the needs of those with JIA. These standards are based upon the experiences and preferences of children and young people with JIA and on evidence of good practice. Standard two aimed to address the issue of a lengthy route to diagnosis and suggested a period of six weeks from onset of symptoms to referral to a specialist rheumatology team. Delayed diagnosis is frequently reported by families (Davies et al. 2010) and can have an adverse effect on both health outcomes and parental/child satisfaction with care.

Therefore in JIA the quicker a diagnosis is made then the speedier treatment can be delivered reducing the chances of joint damage and disability. In JSLE, the experiences of young people in the period leading up to diagnosis can also be particularly difficult and the expertise of specialist teams is necessary to distinguish between lupus and symptoms commonly experienced by children and adolescents. Improved access to care could help reduce the impact and burden of disease for young people by facilitating earlier diagnosis and access to treatment. The focus of this critical exploration will centre upon the period from when the young person experiences the first symptoms of their illness and conclude when a diagnosis is established. The challenge to specialist rheumatology teams providing holistic care to young people with JSLE is considerable. The consequences of getting the diagnosis wrong can result in morbidity and even worse, death, in a young person with lupus. This study is important as the participation of children in research related to their disease is crucial to enhance the understanding of issues that are relevant to them, improve outcomes and facilitate the development of standards of care in JSLE.

## CHAPTER II: LITERATURE REVIEW

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### Introduction

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The impact of being diagnosed with a rare, multisystem, severe, chronic and in some cases life threatening condition is incomprehensible to most people. In Juvenile Systemic Lupus Erythematosus (JSLE) the person receiving the diagnosis is a child or young person whose life will be changed by the diagnosis of JSLE, a disease process which at present, is incurable. Whilst evidence exists on making a diagnosis, disease management and treatments, this literature review aims to identify evidence relating to the experiences of the child or young person before and during diagnosis of JSLE. The intention was to explore and gain insight into what it is like to be a young person developing and receiving a diagnosis of JSLE, thus any literature pertaining to adult disease was not included.

A systematic search was undertaken in January 2014 using the key words: systemic lupus erythematosus; child; adolescent; diagnosis and experiences; searching the databases of Cumulative Index of Nursing and Allied Health Literature (CINAHL), Excerpta Medica Database (EMBASE), Medline, PsycINFO and British Nursing Index see Table I. The databases were searched between 1993 and 2013.

**Table I: Search strategy**

Terms used and structure of search
1. (systemic AND lupus AND erythematosus).ti,ab
2. exp LUPUS ERYTHEMATOSUS, SYSTEMIC/
3. 1 OR 2
4. 3 [Limit to: (Age Groups Child~ 6-12 years or Adolescent~ 13-18 years)]
5. diagnosis.ti,ab [Limit to: Publication Year 1993-2013 and (Age Groups Child~ 6-12 years or Adolescent~ 13-18 years)]
6. 4 AND 5 [Limit to: (Age Groups Child~ 6-12 years or Adolescent~ 13-18 years) and Publication Year 1993-2013]
7. experienc*.ti,ab [Limit to: Publication Year 1993-2013 and (Age Groups Child~ 6-12 years or Adolescent~ 13-18 years)]
8. qualitative.ti,ab [Limit to: Publication Year 1993-2013 and (Age Groups Child~ 6-12 years or Adolescent~ 13-18 years)]
9. (systemic AND lupus AND er*thematosus).ti,ab [Limit to: Publication Year 1993-2013 and (Age Groups Child~ 6-12 years or Adolescent~ 13-18 years)]
10. exp LUPUS ERYTHEMATOSUS, SYSTEMIC/DI, PF [DI=Diagnosis, PF=Psychosocial Factors]
11. exp LUPUS ERYTHEMATOSUS, SYSTEMIC/DI [DI=Diagnosis]
12. 4 AND 11 [Limit to: (Age Groups Child~ 6-12 years or Adolescent~ 13-18 years)]

This search was unsuccessful in yielding any papers therefore an integrative review framework was adopted as this has been recognised as the broadest type of review, allowing the inclusion of experimental and non-experimental research, and can also include theoretical and empirical literature in order to enhance knowledge and understanding of a particular phenomenon (Whittemore & Knafel 2005). Thus by widening the review process different types of sources can be selected. The integrative review framework I chose to utilise has five stages: problem identification, literature search, data evaluation, data analysis and presentation (Whittemore 2005) and these five stages provide structure for this chapter.

### Problem Identification.

The aims of the review as stated in the introduction were to:

- Identify evidence relating to the experiences of the child or young person before and during diagnosis of JSLE.
- Explore and gain insight into what it is like to be a young person developing and receiving a diagnosis of JSLE- ( therefore any literature pertaining to adult disease not included)

This stage involved further development, clarification and simplification of these aims to guide the varied sampling of sources which are particular to an integrative review. This resulted in the purpose of the review being driven by two questions:

1. How is a diagnosis of Lupus in a young person made?
2. How are the lives of young people affected by lupus?

## Literature Search

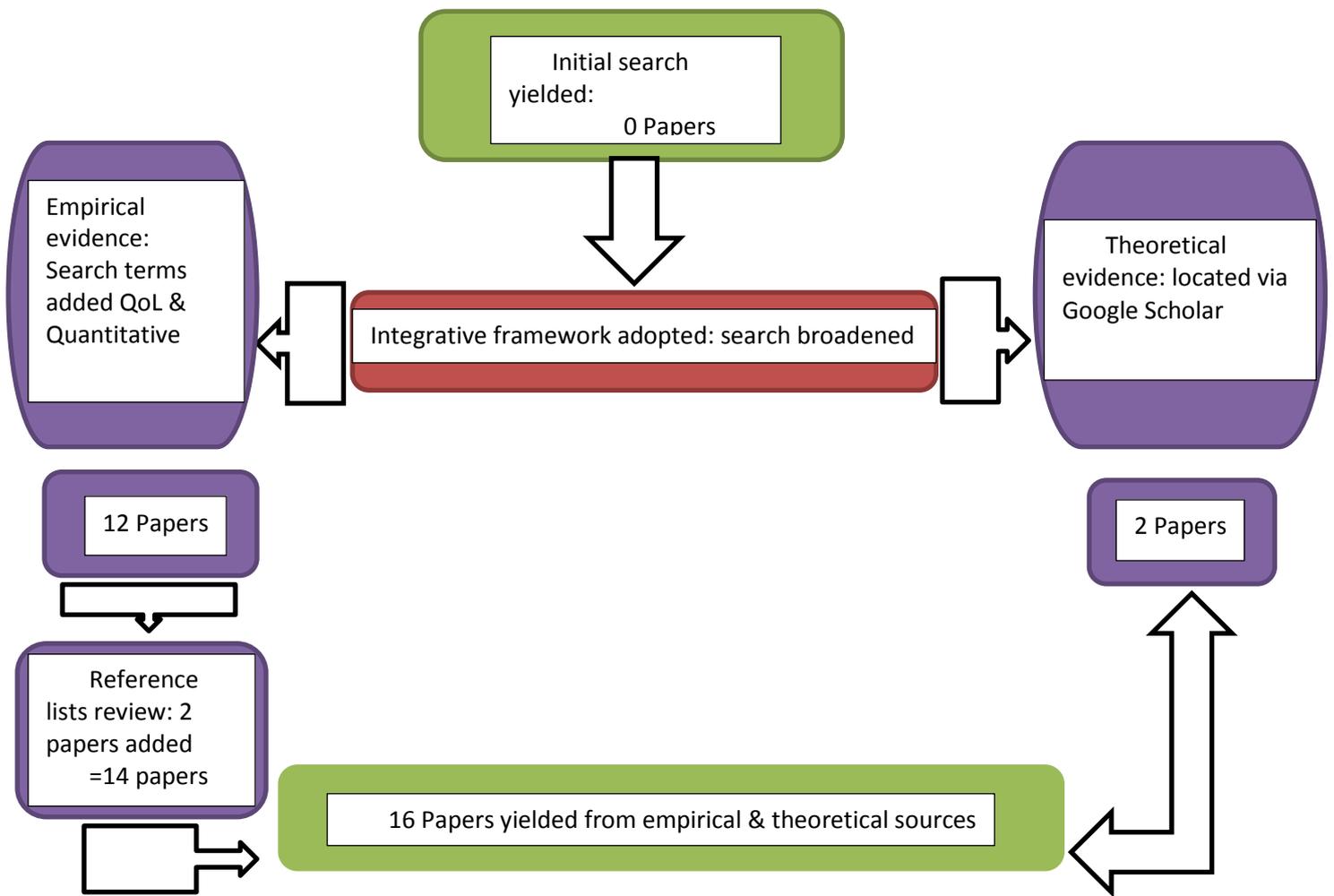
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A purposive approach to sampling was necessary to identify appropriate sources. To review literature on the first question the search was limited to primary sources from the American College of Rheumatology as these criteria were used in the diagnosis of all the eligible young people at the tertiary centre and these two key papers were located through using Google Scholar. Next, the initial database search was rerun (Table 1) with additions of the key term, quality of life and quantitative research papers included. This yielded 12 papers, was augmented by review of the reference lists of all papers, and a further two sources were selected (see Figure 2 for literature search process). Due to the paucity of literature the researcher made the decision to include conference abstracts (poster presentations) published as journal supplements. These were only included when the quality of the underlying research or evidence could be reviewed. The paucity of literature reflects the fact that the research in this area is still in its infancy.

Quality of life (QoL) is a widely applied concept. In this study it is deemed to be a concept used by researchers to determine the impact a chronic disease may have upon an individual in terms of mental, physical and social functioning including perceptions of health, fitness and wellbeing (Bowling & Windsor 2001). By addressing QoL the researcher aimed to capture literature on how JSLE affected wider aspects of a young persons' life. However, it is important to recognise that most studies into health related quality of life for young people with JSLE were undertaken primarily as an outcome measure in clinical trials. This therefore means that such studies are unlikely to capture every aspect of personal experience. JSLE is heterogeneous in nature and diagnosis coincides with

young people going through different developmental stages (child-adolescent-young adult); this further compounds difficulties in the use of measurement tools to determine QoL (Moorthy et al. 2004). However QoL studies can offer insight into how young people’s lives are affected by JSLE, providing valuable background, and for this purpose warrant inclusion in this literature review. Studies that focused solely on instrument development designs and validation of tools to measure QoL were excluded from the review. However during the data analysis process brief information is provided on QoL tools and disease monitoring tools to aid the analytical process.

By focusing the review the literature search retrieved two theoretical sources pertaining to diagnosis of lupus and 14 empirical sources which focused on QoL in young people with lupus as potentially relevant.



**Figure 2: The literature search process**

## Data Evaluation

The final sample for this integrative review included theoretical and empirical reports. Due to the diverse presentation of primary sources, data were evaluated using two criteria relevant to this review: methodological rigour and data relevance using a two-point scale (high or low). An adaption of a Critical Appraisal Skills Programme (CASP) Checklist was used to evaluate rigour and relevance by considering the following issues:

- Are the findings and methodology of the paper valid?
- What are the findings?
- Will the findings help/are they relevant to the aims?

Due to the fact that there were so few studies available to review, after much thought and discussion, it was decided not to exclude any paper based upon this rating. However, less reliance was placed on those with low rigour and relevance and these papers naturally contributed less in the analysis stage (See Table II showing data evaluation process).

**Table II: The data evaluation of the reviewed papers (n=16) showing High and Low Scores.**

<b>Author (Year)</b>	<b>Aim</b>	<b>Sample size</b>	<b>Methods &amp; Rigour score</b>	<b>Key findings and relevance score</b>
<b>Tan et al. (1982)</b>	To describe the revised criteria for the classification of Systemic Lupus Erythematosus	Not applicable	Theoretical descriptive  LOW	Classification system for systemic lupus which has since been updated.  LOW
<b>Hochberg et al. (1997)</b>	To describe the updating of the American College of Rheumatology revised criteria for classification of systemic lupus erythematosus	Not applicable	Theoretical descriptive  HIGH	Updated classification system devised to determine a diagnosis of lupus for use in clinical trials.  HIGH
<b>Moorthy et al. (2004)</b>	To investigate if and in what ways JSLE impacted on QoL as perceived by child and/or parent.	21 Young People (YP) with JSLE, 16 parents	Qualitative prospective single centre using semi-structured technique. Grounded theory used in analysis.  HIGH	Both groups revealed that lupus significantly affected QoL even though disease activity in the population low during the study period. Only qualitative study.  HIGH
<b>Ruperto et al. (2004)</b>	To evaluate the relationship between the health related QoL in different health and the presence of disease	297 YP with JSLE Control groups:	Cross sectional comparative multi centre with children from	Decline in QoL with progressive disease activity. Especially in central nervous, renal &

	activity and damage in the organs that can be affected by JSLE	JIA-649, healthy children 1,333	Italy, Greece, US, Mexico and Japan	musculoskeletal systems. Accumulated damage impacted on physical health and family functioning but did not appear to effect mental health.
			HIGH	HIGH
<b>Moorthy et al. (2005a)</b>	To assess the relationship with JSLE disease activity on both QoL and physical function	24 YP with JSLE, 19 parents	Cross sectional study carried out in 1 tertiary centre for rheumatology in the US.	Neither QoL or physical function strongly correlated with JSLE activity and both need to be considered in order to obtain a comprehensive evaluation of JSLE's impact on children.
			HIGH	HIGH
<b>Moorthy et al. (2005b)</b>	To discuss multidimensional aspects and bio-psychosocial implications of JSLE and factors complicating QoL assessment	Not applicable	Review	Highlights need for a paediatric specific QoL tool.
			LOW	LOW
<b>Moorthy et al. (2007b)</b>	To discuss challenges of evaluating QoL, different scales and the development of a paediatric specific QoL tool.	Not applicable	Review	Understanding QoL issues can help mitigate distress secondary to medical and psychosocial implications of JSLE. Need for a specific tool to measure QoL .
			LOW	LOW
<b>Demirka ya et al. (2008)</b>	To evaluate the neuropsychiatric involvement in young people with JSLE	20 YP with JSLE 20 healthy controls 20 YP with chronic disease	Cross sectional study in one centre in Turkey	YP with JSLE were significantly more depressed when compared to healthy controls and diseased controls. School performance declined in those with moderate to severe depression. In YP with JSLE who displayed mood disorders neither there family nor doctor had detected signs.
			HIGH	HIGH
<b>Brunner et al. (2009)</b>	To compare disease course in JSLE and it's relationship to QoL	98 YP with JSLE	Longitudinal study multi centre study in US.	QoL reduced with increasing disease activity in general, musculoskeletal, neurological and vascular systems. But not in mucocutaneous, renal, and cardiovascular systems. YP displayed social scores comparable with healthy children.
			HIGH	HIGH
<b>Moorthy et al. (2009)</b>	To assess the relationship between QoL and SLE activity and damage in children over time	68 YP with JSLE, 61 parents	Prospective longitudinal multi centre study in US using SMILEY(QoL tool	Parent scores correlated with increases in disease damage but not with a change in disease activity.YP scores

			devised for JSLE)  HIGH	correlated with an increase in damage and were also responsive to changes in disease activity.  HIGH
<b>Moorthy et al. (2010)</b>	To examine YP's and parents perception of impact of JSLE on school, the relationship between YP and parent reports, and the relationship between QoL and YP satisfaction with school performance including interest, memory and concentration.	41 YP with JSLE, 32 parents	Prospective cross sectional study in two centres in US  HIGH	Both parents and YP felt that JSLE had a significant impact on school related issues and perceived JSLE reduced participation in school life . 50% missed school due to not feeling well, 74% missed school due to medical appointments.  HIGH
<b>Sousa &amp; Guedes (2011) poster</b>	To assess the health related QoL in JSLE in using PedsQL3.0 rheumatology module	6 YP with JSLE	Cross sectional study in paediatric department in Portugal  LOW	Highlighted need for improving psychological support for YP and families.  HIGH
<b>Louthrenoo et al. (2012)</b>	To assess the emotional and behavioural problems in childhood lupus in a nephrology clinic in Thailand	40 YP with JSLE, 40 healthy controls	Cross sectional study in one centre in Thailand  HIGH	YP with JSLE displayed evidence of psychosocial adjustment during low disease activity and were not found to be at risk of psychosocial dysfunction.  HIGH
<b>Moorthy et al. (2012)</b>	To provide an update on the assessment of QoL in children with JSLE	Not applicable	Review  LOW	QoL relevant to YP with JSLE, generic and disease specific tools exist. QoL should be examined in different geographic/ethnic populations. There is a need to improve understanding of issues that significantly impact on QoL and devise interventions to improve the QoL in YP with JSLE.  HIGH
<b>Tuck et al. (2012) poster</b>	To assess the QoL in recently diagnosed YP with JSLE	20 YP with JSLE, 20 parents	Cross sectional study in one centre in US  HIGH	Social functioning scores using the PedsQL in recently diagnosed YP with JSLE were relatively less affected when compared to physical, emotional and school functioning.  HIGH
<b>Moorthy et al. (2013) poster</b>	To examine the correlation between YP and parent health related QoL scores	267 YP with JSLE, & their parents	Cross sectional study in US and Latin America.  HIGH	Parents and YP have different perspectives about the impact on QoL. Parents tend to over estimate impact. HIGH

The reviewed evidence was analysed to provide context, richness and depth into the exploration of the experiences of young people during diagnosis. The discussion centres upon medical definitions of making a diagnosis JSLE. The American College of Rheumatology (ACR) 1982 revised classification criteria (Tan et al. 1982) and updated by Hochberg et al. (1997) (see Table 3) was used for the purpose of identifying patients in clinical studies and a person is said to have systemic lupus erythematosus if any four or more of the eleven criteria are present, serially, or simultaneously during any period of observation. Using these criteria emphasis is placed upon classification rather than diagnosis. In the case of a young person, nevertheless, making a diagnosis rests ultimately with a consultant paediatric rheumatologist experienced in the assessment and management of lupus in children and young people as a single diagnostic test does not exist. Differing from the disease diagnosed in adults, young people can benefit from an earlier diagnosis of the disease and, as in other rheumatic conditions, the earlier the disease is detected the earlier treatment can be initiated thus preventing irreversible damage from inflammation occurring. It is for this reason that paediatric rheumatologists may classify a young person as having lupus if they have only two or more ACR criteria due to the likelihood of probable or evolving disease in the paediatric population.

**Table III Classification Criteria for Systemic Lupus Erythematosus** (adapted from American College of Rheumatology, 1997)

Symptom	Description
<b>Malar rash</b>	A butterfly –shaped rash over the cheeks and bridge of the nose.
<b>Discoid rash</b>	Red raised patches of skin with scaling and plugging of hair follicles
<b>Photosensitivity</b>	Skin rash as a result of an unusual reaction to sunlight
<b>Mucosal ulcers</b>	Small ulcers usually painless occurring in mucosal lining of nose or mouth
<b>Arthritis</b>	(non-erosive) involving two or more peripheral joints, characterised by swelling
<b>Serositis</b>	Inflammation of the serosal surfaces: pleuritis, pericarditis
<b>Renal disorder</b>	Persistent protein and cellular casts in the urine
<b>Neurological disorder</b>	Seizures or psychosis
<b>Haematological disorder</b>	Haemolytic anaemia, Leukopenia, Lymphopenia, Thrombocytopenia
<b>Immunological disorder</b>	Anti-DNA; Anti-Sm ; Positive finding of antiphospholipid antibodies
<b>Antinuclear antibody</b>	An abnormal titre of antinuclear antibody

Although the ACR criteria are widely used in the diagnosis of SLE in 2012 the Systemic Lupus Collaborating Clinics (SLICC) group recently developed and validated a new set of alternative criteria. These new criteria have yet to be validated in young people but its recent application using data from the UK JSLE cohort increased the numbers of young people meeting a definite diagnosis of lupus (Lloyd et al. 2013). This suggests the SLICC criteria is potentially more sensitive than the ACR criteria for diagnosing lupus in younger patients and the use of SLICC criteria may enable earlier diagnosis of lupus. This is of particular importance in young people who often have more aggressive disease and would benefit from early treatment but further studies are needed to assess the application of the SLICC criteria for classification of lupus. All young people in this study were diagnosed according to the ACR criteria.

Advances in research and the development of new drugs and treatments have demanded outcome measures to determine their success or failure. Since 2004 there has been an increase in both the development and use of tools validated to capture and report on critical determinants of outcome utilised in JSLE (Ravelli et al. 2005). Outcome measures are variables representative of disease or person specific components which when captured together can track improvement, deterioration or lack of change in the condition of the young person. In JSLE the British Isles Lupus Activity Group (BILAG) score is a validated outcome measure (Brunner et al. 1999) and is used to determine lupus activity in eight organ systems or domains. This scoring system highlights the organ or domain affected by disease and also the severity to which that system is compromised. The BILAG is measured and completed by the physician. Another activity score validated for determining disease activity in lupus is the Systemic Lupus Erythematosus Disease Activity Index (SLEDAI) (Bombardier et al. 1992) and again like BILAG scored by the examining physician. In comparison, health related QoL measures are also component tools but completed by the individual thus enabling patient participation in outcome. It is crucial when considering outcome scores to determine who has done the scoring and to reflect on how this may influence scoring. There are a number of tools that have been used in JSLE to measure both QoL and physical function/disability. It remains beyond the scope of this review to critically discuss individual tools but the four main QoL tools that have been used in children and young people with JSLE are briefly discussed.

The Childhood Health Assessment Questionnaire (CHAQ) measures functional health status and was originally validated for use in juvenile arthritis (Singh et al. 1994). It comprises eight categories for assessment such as dressing and grooming. There is also a visual analogue scale for assessment of pain and overall health. Parents of children (< 11 years old) complete the CHAQ and young people above age 11 complete their own CHAQ. It is the most widely used score and has shown responsiveness over time in young people with lupus (Hersh 2011).

The Child Health Questionnaire (CHQ) is designed to measure physical, emotional and social aspects of health and has 14 categories for assessment (Ruperto et al. 2004). Again it has two forms, one for parents and one for children and young people aged 11 and above. It has been shown to be sensitive to changes in disease activity in lupus over time but has not specifically been tested in young people (Hersh 2011).

The Pediatric Quality of Life Inventory-Generic Core Module (PEDSQL-GC) and Pediatric Quality of Life Inventory- Rheumatology Module (PEDSQL-RM) are used in combination. The PEDSQL-GC is a general assessment of health related QoL using four categories of physical functioning, emotional functioning, social functioning and school functioning whereas the PEDSQL-RM is specific to categories pertaining to health related issues in rheumatology. Both PEDSQL-GC and PEDSQL-RM can be completed by a child 5- 18 years or by parent proxy report for ages 2-18 years. Although as in the CHQ they have not been specifically tested in young people with lupus they are quick and easy to complete with the PEDSQL-RM showing particularly sensitivity to changes in health status over time (Hersh 2011).

The Simple Measure of Impact of Lupus Erythematosus in Youngsters (SMILEY) is the only QoL tool specific to lupus in young people. It utilises a novel approach whereupon responses are recorded in the form of different facial expressions and both children with lupus and their parents participated in its development (Moorthy et al. 2007a). However, as a recently developed tool it has yet to be widely adopted and more studies are needed to assess its long-term sensitivity (Hersh 2011).

It was possible to identify the following themes in the literature and the findings are presented under the headings:

- Disease Activity and QoL
- Damage caused by JSLE and QoL
- Social functioning and QoL
- Psychosocial Issues

#### *Disease Activity and Quality of Life*

Researchers have hypothesised that health related QoL is reduced in young people with rheumatic conditions (Varni et al. 2002) including those with a diagnosis of JSLE compared with healthy children due to the burden of living with a chronic multi-system relapsing and remitting condition (Moorthy et al. 2012). Interest has centred round the relationship between QoL and disease variables. Ruperto et al (2004) in a cross sectional comparative multicentre study of 297 children with JSLE from Italy, Greece, US, Mexico and Japan highlighted a decline in QoL with progressive disease activity. Thus it could be assumed during increased disease activity (when a young person's lupus gets worse) that their QoL would be reduced.

However, other studies have failed to reveal a significant correlation between quality of life and disease activity (Houghton et al. 2008, Moorthy et al. 2005a & 2009). In contrast Brunner et al. (2009), in a longitudinal study of 98 children with JSLE from seven paediatric rheumatology centres in the US, compared disease activity in different systems and its relationship to QoL using PedsQL-GC, PedsQL-RM and CHQ. This study revealed a correlation between a reduction in QoL with increasing disease activity in general, musculoskeletal, neurological and vascular systems. This finding was supported by Ruperto et al. (2004) who noted an increase in disease activity in central nervous, renal and musculoskeletal systems resulted in a decline in QoL. Interestingly an increase in

lupus disease activity in mucocutaneous, renal and cardiovascular systems was not found to reduce quality of life (Brunner et al. 2009).

Moorthy et al. (2004) explored issues about the impact of lupus related to QoL in the only study reviewed that adopted a qualitative approach. This research took place in a paediatric rheumatology tertiary centre in the US. This qualitative prospective study used a semi structured technique initiated by a single open ended question related to JSLE which was handed to participants on a piece of paper. Parents or young people chose to either record their responses on the piece of paper or verbally replied to the researcher. Grounded theory was used to identify themes. The aim was to investigate if, and in what ways, lupus impacted on QoL as perceived by the child and parent, rather than using an objective measurement tool.

Moorthy et al. (2004) study was carried out in a population of 21 young people and 16 parents, whose disease activity was defined as low using the SLEDAI score. The young peoples' responses centred round the themes of coping with lupus whilst maintaining control despite their illness. In comparison, parents' answers identified the effort required in coping with their child having lupus as a theme. The young people related these themes to categories of limitations: impact of/on social and family relationships, effect on self, fear of the future and long term goals. Areas highlighted by parents were: psychological, accommodating disease, shifting expectations, social support, worry and medical care. Analysis of the responses of both young people and parents implied that JSLE significantly affected QoL even though disease activity in the population was identified as low during the study period.

#### *Disease Damage and Quality of Life*

Damage can be defined as any non-reversible change not related to active inflammation occurring since the onset of JSLE, ascertained by clinical assessment and present for at least six

months (Ruperto et al. 2004). It is expected that over time damage scores are likely to increase because of the irreversible nature and inherent long term health consequences of damage to major organ systems. The aim of treatment in the management of JSLE is to prevent it occurring. The Systemic Lupus International Collaborating Clinics Damage Index (SDI) (1996) is a scoring system that has been validated to assess accumulated damage caused by lupus and this method was used to measure damage in all papers reviewed. Brunner (2009) found that young people with damage to any organ system had a significantly reduced QoL compared to those with minimal or absent disease damage. However, the only score not increased by disease damage was the psychosocial summary score. Increased damage was shown to limit parents' free time and lead to disruption of family activities. This finding contrasted with scores for disease activity where a greater impact was seen on parent time but also resulted in increased parental emotional distress.

Further to this Ruperto et al. (2004) found accumulated damage impacted on physical health and family life functioning but did not appear to have an effect on mental health. Ruperto et al. (2004) postulated this may be due to the emotional/psychological ability of the individual to adapt or respond to limitations caused by disease. The ability of young people with lupus to adapt to the stressful events imposed by their illness was also recognised by Louthrenoo et al. (2012). The objective of this cross-sectional study of 40 young people with lupus and 40 healthy controls was to assess the emotional and behavioural problems in childhood lupus in a nephrology clinic at Chiang Mai University Hospital in Thailand. Parents of both groups were interviewed and asked to identify any learning, emotional and other difficulties including economic or family problems that their child may have experienced.

In addition parents completed the Child Behaviour Checklist (Achenbach & Ruffle 2000) and the young people were given the Children's Depression Inventory (Kovacs 1985) and the Multidimensional Anxiety Scale for Children (March et al 1997). The research was conducted in

young people assessed as having low disease activity using the SLEDAI (score 0-1). The results revealed that young people with lupus displayed evidence of psychosocial adjustment towards their illness during periods of low disease activity and they were not found to be at increased risk of psychosocial dysfunction. This indicates that the young people studied were able to manage physical restrictions placed on them by lupus (in the case of irreversible damage) and may overtime adapt without this impacting on mental health.

Moorthy et al. (2009) in a prospective longitudinal study of 68 young people with JSLE and 61 parents from five paediatric rheumatology centres in the US compared parallel scores at baseline (first appointment within the study period) and a follow up appointment using SMILEY (the QoL tool specific to lupus in young people). The results illustrated that parent scores correlated with changes in disease damage but did not show a correlation with a change in disease activity. Therefore it has been suggested that parents showed an increased awareness of the long-term effects and ongoing disruption to their child's life caused by lupus but are less sensitive to minor changes in disease activity. Correspondingly, child scores correlated with an increase in damage assessment and were also responsive to changes in disease activity.

#### *Social Functioning and Quality of Life*

Social functioning has been defined as an index of children's interest and performance in a variety of areas including participation at school, social and sporting activities, even how easily a young person makes friends (Adams et al. 2002). All of which can be considered formative in the transition of a young person into an adult. A chronic illness has been shown to have a detrimental impact on the ability of a young person to participate in social interactions due to illness perceived barriers by parents who restrict the young person involvement (Wagner et al. 2003). However, Brunner et al. (2009) found the young people with lupus studied displayed social scores comparable with those of healthy children. Interestingly 20 adolescents recently diagnosed with JSLE (within 18

months of diagnosis) revealed social functioning scores using the PedsQL that were relatively less affected when compared to physical, emotional and school functioning domains (Tuck et al. 2012). The timing of scoring could therefore be significant as during the first few months following diagnosis young people may be unaware of social limitations placed upon them by lupus (perhaps because they are too poorly to want to participate in social activities). However, as the young person moves further on their journey from diagnosis to management of their condition and start to feel better they may become more aware of social restrictions placed upon them by hospital admissions, treatment and symptoms such as fatigue.

Further to this Houghton et al. (2008) in a cross sectional study of fitness, fatigue, disease activity and quality of life in 15 young people with lupus aged between 12 and 19 years of age found no significant relationship between the four variables but results did suggest that the young people discontinued their participation in sport or physical activity around the time of diagnosis. Fatigue was noted to be a significant symptom of the young people in this study that restricted participation in activities. Similarly Moorthy et al. (2010), in a prospective cross sectional study of two rheumatology practices in the United States of 41 young people with JSLE and 32 parent parallel reports using the SMILEY, found both parents and young people felt lupus had a significant impact on school related issues. 50% of young people responded that they missed school due to not feeling well and 74% because of seeking medical attention. This highlights the significant impact lupus can have upon self-reported school issues and also the perceived ability to participate in school life.

### *Psychosocial Issues*

Adolescence is a period defined by adjustments to change albeit physical, mental or social. Adjustment and responsiveness to such changes are personal and often attributed to a nature versus nurture schemata. During adjustment to such changes a young person with a chronic illness can be at an increased risk of poor mental health however evidence shows that they can emerge

from this period with a positive outlook and display resilience to the challenges presented by their illness (Olsson et al. 2003, Noll et al. 2000).

Specifically in JSLE the clinical spectrum of disease may include neuropsychiatric manifestations including mood disorders, psychoses and cognitive deficit. Therein the young person with JSLE could be subject to a threefold challenge: adolescence, psychosocial adjustment to chronic illness compounded by symptoms of neuropsychiatric lupus. Demirkaya et al. (2008) was a cross sectional study aimed at evaluating the neuropsychiatric involvement in twenty JSLE patients compared with 20 healthy adolescent controls and 20 adolescents with chronic disease. This study, which was carried out in one centre for paediatric rheumatology and nephrology in Turkey, found that the young people with lupus were significantly more depressed when compared to both healthy and diseased controls. Depression was assessed using the Symptom Distress Checklist, a questionnaire which was developed to determine psychological disorders and measure severity (Derogatis 2010) and has been used previously in rheumatology patients (Parker et al. 1990). Significantly all of the nine JSLE patients identified as having moderate to severe depression reported a decline in school performance. Further to this, 11 (55%) of the JSLE patients displayed neurological signs and symptoms. Of those JSLE patients displaying mood disorders neither family nor physician had detected signs in the young person.

Recognition of signs and symptoms of disease are challenging for the health professionals, the family and the young person with lupus. Louthrenoo et al. (2012), as previously described, assessed young people with lupus during disease remission and concluded in the population studied that when disease was inactive the young people were not found to be at risk of psychosocial problems. In contrast, Demirkaya et al. (2008) commented that the prevalence of neuropsychiatric disorders, and in particular depression, did not correlate with increased disease activity. The challenges

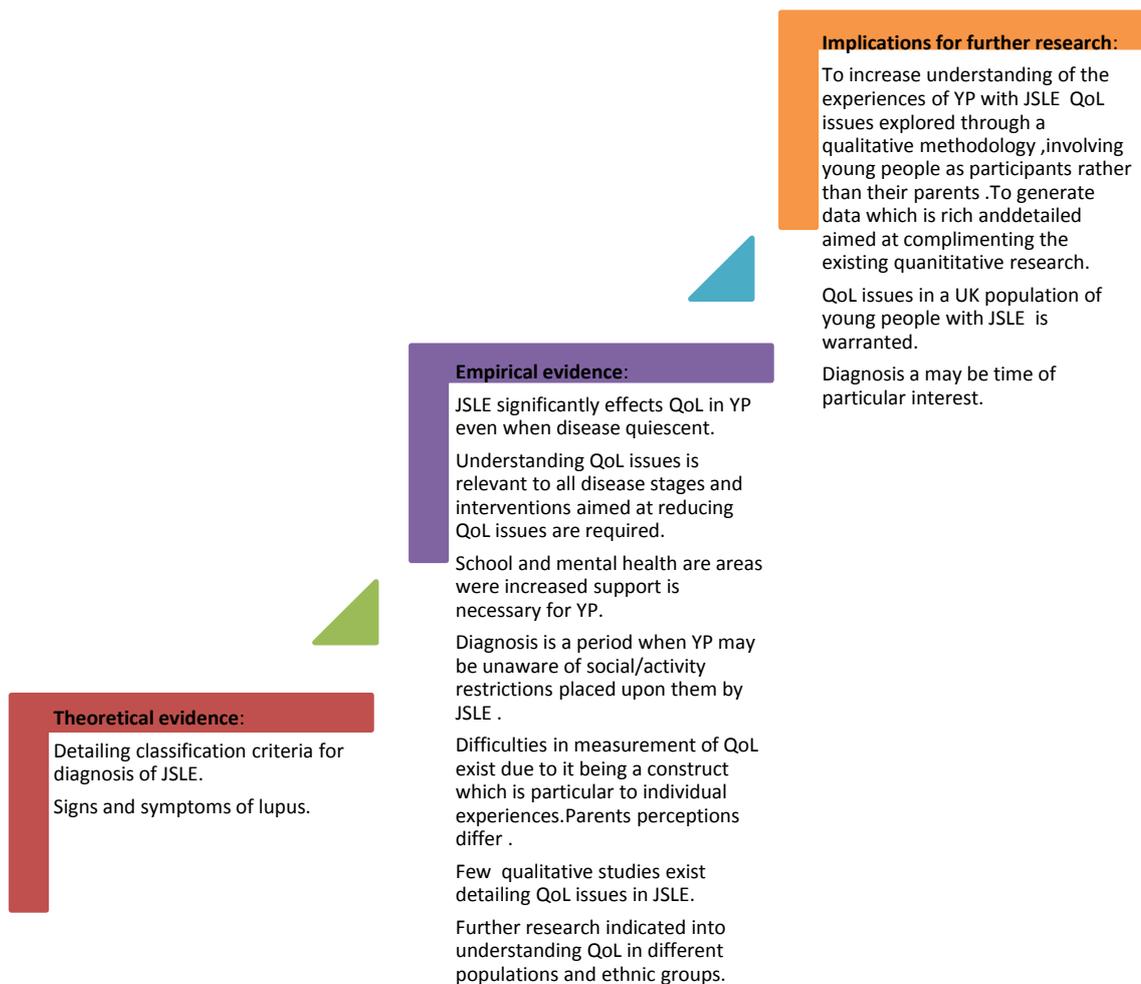
presented by the identification of neuropsychiatric lupus, chronic illness and adolescence, highlight the need for thorough psychological evaluation of young people with JSLE as part of routine care.

## Presentation of Results

In this section the results of the synthesis of evidence through the integrative review process is displayed in diagrammatic format (see Figure 3). The results of this integrative review demonstrate a logical chain (searching, selecting and analysing the papers) and the subsequent integration of evidence that attempts to demonstrate a clear understanding of the literature and existing evidence related to the diagnosis and the experiences of young people with JSLE. The implications for further research are emphasised and the methodological limitations of the integrative review process are discussed and supported by conclusions. The integrative review has provided a solid context and justification for the need to undertake the study.

### *Methodological Limitations of the Review*

Due to the initial search failing to reveal any directly relevant studies into experiences of young people with JSLE it was necessary to conduct an integrative review to enable a broad approach to generation and synthesis of evidence. The inclusion of QoL studies revealed areas and ways young people may be affected by their diagnosis of JSLE but the evidence was limited by the quantitative framework within which they were conducted (with the exception of one qualitative study). The information gathered is subject to closed questioning inherent in QoL tools rather than open ended questioning whereupon a young person may choose their own words and expressions to reflect and capture their experiences of how they feel their life and its quality is affected by JSLE. However, JSLE is a disease of young people and different from adult onset disease. Therefore the inclusion of QoL studies was justified as pertinent to young people, rather than evidence generated from adult qualitative studies. Evidence generated from adult experiences is relevant to a disease present in adulthood which is different from a disease experienced by young people with JSLE. To use such evidence would be to assume similarities and differences to adult experiences rather than focus solely on the experiences of young people with JSLE which was the aim of this review.



**Figure 3: Showing summary points of the synthesis of theoretical and empirical evidence and the implications for future research**

## Conclusions

The evidence presented suggests that the heterogeneity of JSLE extends further than the disease process itself and can also affect QoL in relation to disease activity, disease damage, social functioning and psychosocial issues. Despite their potential strengths in gaining objective data across populations, caution must be exerted when considering the findings from QoL tools as they may not capture a comprehensive analysis of individual responses and/or experiences. With so little data

reflecting the experiences of young people with JSLE, qualitative research provides a framework to facilitate this exploration, and this exploration is warranted particularly during the pathway to diagnosis of JSLE.

## CHAPTER III: METHODOLOGY

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### Introduction

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An exploratory study using a qualitative methods approach was undertaken involving individual in-depth interviews with eight young people diagnosed with lupus in one UK tertiary centre for rheumatology. Ethical approval was obtained via the National Research Ethics Service (NRES), and the study was also approved by Alder Hey Research Business Unit and by the BuSH Ethics Committee at the University of Central Lancashire. The researcher approached the study drawing on key principles from critical realism. Whilst critical realism covers a broad range of approaches (Trochim 2006) there is a basic acceptance that 'there is a real world out there' (Easton 2010, p118) and that the 'production of any kind of knowledge is a social practice' (Easton 2010, p120). Critical realists can use both quantitative and qualitative methods in their research. For this study, the researcher chose to use qualitative interviews as the method. The goal was to endeavour to reveal the reality of the experiences a young person with JSLE may encounter from their perspective. As a critical realist, the researcher was aware that however robust the research process was it would be never achieve an absolute all-encompassing truth. However, by collating and critically analysing the experiences of young people the researcher aimed to ascertain a view of their reality about their experiences from symptom onset to diagnosis of JSLE.

### Aims of the study

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The aims of the study were to:

1. Describe the journey from onset of symptoms to diagnosis in JSLE from a childhood/adolescent perspective by exploring the stories that young people tell about their experiences of this period in one tertiary centre.
2. Ascertain key points, if any, in the journey to diagnosis to generate deeper insight into access to care for young people with JSLE.

## Target Population

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The study engaged young people undergoing care for JSLE and who were already participating in the UK JSLE Cohort Study at the researcher's own tertiary centre. This constituted a target population of 23 young people. The median age at diagnosis for JSLE patients has been demonstrated as 12.6 years (Watson et al 2012). All the young people who participated were aged between 12 and 19 years and only one was male. From the target population of 23 young people only nine satisfied the inclusion and exclusion criteria (see below). One young person declined participation. The 14 young people who failed to meet the inclusion and exclusion criteria were diagnosed with JSLE prior to 1<sup>st</sup> January 2010. With the exception of one young person all eight were willing to participate in this study and all were involved and supportive of research through the UK JSLE Cohort Study.

### Inclusion criteria:

1. Participating in the UK JSLE Cohort Study.
2. Young people diagnosed with JSLE or evolving JSLE within the last two years (from 1st January 2010) were invited to participate as it is felt this would generate data that reflected contemporary service delivery and standards of care. **Nine young people met the criteria.**
3. Any young people newly diagnosed within the period of data collection were also invited to participate. One young person did not want to participate due to the fact that they were newly diagnosed and did not feel able whilst still in the initial stages of treatment.
4. Due to the small numbers involved it was felt that for any of the young people and parents who were non-English speaking an effort would be made to make use of an interpreter to enable them to participate. One parent was non-English speaking and appropriate measures were taken during the informed consent process.

Exclusion criteria:

1. Diagnosis more than two years ago (before 1st January 2010).
2. No longer receiving care at the tertiary centre.

## Recruitment and Consent

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The study recruited young people identified by their consultant as suitable to approach who were already participating in the JSLE cohort study, cared for at the tertiary centre and who met the inclusion/exclusion criteria. Detailed parent and age-appropriate young person study information sheets were distributed by post two weeks before a clinic visit to ensure adequate time for reading (see Appendix 1). At the clinic visit, an opportunity was given for questions to be asked by both the young people and their parents/carers. Copies of the study information were also made available at the clinic visit and any supplementary information was provided about the study as appropriate, and then the young person was invited to take part. Written informed consent or written parental consent and assent (see Appendix 2) from young people who were under 16 years of age was sought in accordance with good ethical practice. The interviews then took place after the clinic visit at the hospital in the Clinical Research Facility (CRF). The CRF is away from the busy clinic environment and has a quiet room. This was chosen as a suitable place as the young people visit the CRF regularly due to their participation in the JSLE cohort study and are therefore familiar with the surroundings. There is a purpose built comfortable quiet room that provided an excellent venue for the interviews to take place that did not involve disruption to the usual clinic routine for the young people as they visit the CFR after every clinic visit.

## Methods

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An exploratory study using a qualitative methods approach was undertaken which involved individual in-depth interviews with eight young people. Two of the young people were approached after data analysis to review the key points generated from the interviews.

### In-depth interviews

Data were collected by using in-depth interviews with eight young people and during these interviews the young people's experiences from first symptoms to diagnosis were explored. In-depth interviews allow detailed investigation into an individual's perspective, provide personal context to the research phenomenon and exploration of sensitive issues (DiCicco-Bloom & Crabtree 2006, Lewis 2003, Pope & Mays 1995). Individual in-depth interviews were used rather than group interviews providing each young person with the opportunity to develop and draw upon their own experiences. Due to my knowledge and experience working with each of the young participants I felt a group environment might have prevented and intimidated some of the young people (who I knew well) expressing themselves fully. The advantage of previously working with the young people enhanced their engagement in the interview process. However, this might be counteracted by a reluctance to fully explore their experiences especially from those young people with whom I was involved from initial diagnosis. I supported and encouraged the young people to tell the story of their own 'journey to diagnosis' using open questioning based on both my nursing experience and my research skills and knowledge.

An interview schedule was developed (see Appendix 3) under the guidance and support of the supervisory team and staff within the Children's Nursing Research Unit at Alder Hey NHSFT and advice sought from a small number of young people who were participating in the UK JSLE Cohort

Study when they attended Alder Hey as part of their routine care. The young people commented on topics that they might discuss relating to their experiences of diagnosis. I worked upon an interview schedule as it created a framework, as a novice researcher, to undertake the interviews as I was worried that trying to undertake an interview with no structure would be too daunting and I might miss a key area of questioning. However, I did not intend that the schedule would restrict my interaction and engagement with the young people and intended to be flexible in my approach. However, in practice I conducted the interviews without reference to the schedule. In fact, in an effort to ensure listening and appropriate responses to the young people, I found it important to maintain eye contact and focus. Reference to an interview schedule would have distracted me and therefore I chose not to refer to it. The process of preparing the schedule proved beneficial in highlighting topic areas useful as prompts to facilitate the discussion and it was these topics that I drew upon.

Before the interview started I introduced myself, gained consent and talked to the young people about how they could stop the interview at any time. I showed them the recorder and explained what would happen. I reminded them that I was a researcher doing a research study and not their 'nurse' during the interview process (I did not wear my uniform to facilitate this distinction). Confidentiality issues were addressed, I confirmed they were happy to be interviewed without their parent present and we made arrangements for their parent to come to collect them (one young person who was eleven preferred her parent to be present). Once we were both comfortable the interview began.

During the interview I followed the lead of the young person and used prompts such as: "and then what happened" to encourage a comprehensive and detailed account. The interviews were digitally recorded, encrypted and then coded by the use of a number system. A record of the

number that corresponds to the demographic details of each young person was kept separately from the transcripts and digital files. The interviews took place at the tertiary centre, in a private quiet room in the Clinical Research Facility (CRF) and lasted for no longer than one hour. A time was chosen that was convenient to participants and coincided with a clinic appointment, avoiding extra travel and parking charges. Data collection took place from July 2013 to March 2014.

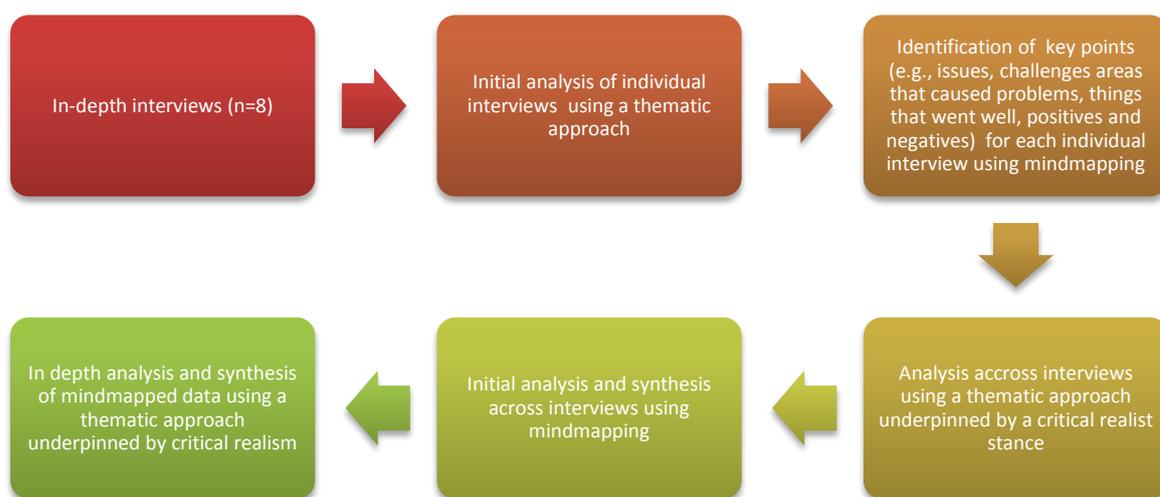
I interviewed each young person individually with the exception of one (the youngest participant) who was interviewed with a parent present. The young people were given the choice if they would prefer the interview to be conducted on their own or with their parent/carer depending on the individual young person and their parent's/carer's preference. Evidence has shown that the presence of parents at interviews can both help and/or limit the contribution of children aged six to thirteen years and the researcher should adopt a reflexive approach to consider and then report the effects of adult influences (Gardner & Randall 2010). However, for this study although the majority of eligible young people were over thirteen for the young person who chose to have a parent present it was crucial that I was aware of this issue and I reported any positive or negative influence that enhanced the young person's view or prevented it being expressed. I found that I had to on occasion, to re-direct questioning towards the young person after the parent had answered. However I felt this enhanced data collection as I was able to listen to both the young person and parent's perspective. Only a few notes were taken during interviews in order to enable me to focus my attention fully upon the young person and facilitate an in-depth questioning approach (Legard et.al 2003). Following each interview, however, I made field notes about key elements of the interview based upon my reflections to provide a supporting context to the interviews. I also used these notes as a guide, being a novice researcher, to highlight things that went well, things that I felt did not go as well and improvements that I could make in the future. It was helpful to reflect upon how the young person and I interacted, whether I felt they were nervous, able or reluctant to talk and I was able to draw on these reflections during data analysis.

I transcribed the interviews as part of the familiarisation process with the data set and this helped facilitate initial analysis. All interview recordings and study data were stored in accordance with the tertiary centre’s Trust’s data protection requirements.

## Data Analysis

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As this study aimed to describe the journey through diagnosis of SLE from the perspective of the young people who have experienced it, a thematic approach to analysis was used (see Figure 2). Thorne (2000) notes that it is through using analytic processes that the main themes within the accounts people give about their lives can be detected and researchers can discover how people understand and make sense of their lives.



**Figure 4: The data analysis process**

The data generated from the interview transcripts were subject to thematic analysis. This approach has been recognised as a research tool which can provide a rich and flexible account of the data, whilst also highlighting any similarities and differences (Braun & Clarke 2006). The epistemological stance adopted was critical realism where the analysis sought to remain close to the experiences from the perspective of the young people (Braun & Clarke 2006). Critical realism was utilised as an approach throughout the analysis by seeking to acknowledge the ways the young people interpreted their experiences from their journey from onset of symptoms to diagnosis. The context of this setting for the young people is reflected during the thematic analysis and the researcher has sought to remain focused upon this reality whilst also allowing for a wider and more complex interpretation of individual experiences.

Each interview transcript was read and themes extracted. The aim of the initial analysis was to create a list of broad themes as well as a list of core ideas (e.g., issues, challenges, areas that caused problems, things that went well, positives and negatives); these were then explored using mind mapping. Mind mapping is a process whereby concepts or themes are arranged around a central key word (Buzan 1993), and it has been recognised as a tool to facilitate the data analysis process in qualitative research (Burgess-Allen & Owen-Smith 2010). The transcripts were reviewed and themes extracted for each individual young person according to core ideas generated and then mind mapped for each young person (see Appendix 4).

The interview transcripts were then reviewed for a second time before themes that were relevant across all interviews were identified and synthesised onto further mind maps (see Appendix 5). Finally the transcripts were reviewed for a third time, through in depth analysis and any additional themes identified were then added to the mind maps. Further analysis drawing on a thematic approach were then undertaken by myself with the support of the clinical nursing research

unit at Alder Hey to synthesise mind maps into major and sub themes, organised chronologically to reflect the young people’s ‘journey to diagnosis’ and a meta-theme linked the themes together.

## Evaluation Using CASP

The study was subjected to evaluation by the Critical Appraisal Skills Programme appraisal tool for qualitative research (see below):

The study was subjected to evaluation by the Critical Appraisal Skills Programme appraisal tool for qualitative research (see Table IV):

**Table IV: Evaluation of the Study using CASP**

CASP appraisal statement	Achieved Yes/No	Evidence to support evaluation claim
Was there a clear statement of the aims of the research?	<b>YES</b>	<ul style="list-style-type: none"> <li>• <i>The aims of the research were clearly stated (as below)</i></li> <li>• <i>Describe the journey from onset of symptoms to diagnosis in JSLE from a childhood/adolescent perspective by exploring the stories that young people tell about their experiences of this period in one tertiary centre.</i></li> <li>• <i>Ascertain key points, if any, in the journey to diagnosis to generate deeper insight into access to care for young people with JSLE.</i></li> </ul>
Is a qualitative methodology appropriate?	<b>YES</b>	<ul style="list-style-type: none"> <li>• <i>The literature review identified the gap in qualitative research and the methodology chapter identified the qualitative approach taken</i></li> <li>• <i>Exploratory research into the experiences of young people is best undertaken using a qualitative approach</i></li> </ul>
Was the research design appropriate to address the aims of the research?	<b>YES</b>	<ul style="list-style-type: none"> <li>• <i>This was identified and discussed in detail during the integrative review process and the justification for a qualitative research design was presented in the methodology chapter. Also critique of the approach is identified in the limitations.</i></li> </ul>
Was the recruitment strategy appropriate to the aims of the research?	<b>YES</b>	<ul style="list-style-type: none"> <li>• <i>The recruitment strategy was appropriate given the limitations imposed within a single centre study and the limitations of the study being undertaken by a novice researcher-practitioner for a MSc. The choice of recruitment strategy is explained and</i></li> </ul>

		<i>reasons are discussed for the non-participation of one eligible young person.</i>
Was the data collected in a way that addressed the research issue?	<b>YES</b>	<ul style="list-style-type: none"> <li><i>The choice of interviews was a reasonable decision for this study. In depth interviews are justified as a method of data collection however the setting for the data collection for convenience on the hospital site away from the clinic area might have been improved if the researcher had conducted the interviews in the homes of the young people. Due to the small sample size data saturation was not discussed as it was not expected that it would be an issue.</i></li> </ul>
Has the relationship between the researcher and the participants been adequately considered?	<b>YES</b>	<ul style="list-style-type: none"> <li><i>The researcher identified the potential power dynamics and tensions in her role as a practitioner-researcher and critically examined her role in the research process, clearly stating advantages and disadvantages.</i></li> </ul>
Have ethical issues been taken into consideration?	<b>YES</b>	<ul style="list-style-type: none"> <li><i>All ethical issues were conscientiously considered prior to the start of the study and care was taken throughout the study to protect the children in line with ethics approval and governance requirements.</i></li> </ul>
Were the data analysis sufficiently rigorous?	<b>YES</b>	<ul style="list-style-type: none"> <li><i>Data analysis was as rigorous as possible given the fact that this was a MSc study. The process of thematic analysis was clearly in sufficient depth description to allow the reader to appreciate the rigour. Quotes were used to illustrate and provide an evidence based to the themes presented.</i></li> </ul>
Is there a clear statement of findings?	<b>YES</b>	<ul style="list-style-type: none"> <li><i>The findings are discussed clearly in relation to the aims of the research and linked to the existing published literature and evidence.</i></li> </ul>
How valuable is the research?	<b>YES</b>	<ul style="list-style-type: none"> <li><i>Qualitative research in this area is in its infancy and this study makes a significant contribution towards the experiences of young people with JSLE and may also be considered relevant to young people diagnosed with other rare rheumatic conditions.</i></li> </ul>

## Summary

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In summary I worked with the young people to develop topic areas for discussion during interview and was careful to give the young people space and time to talk about their experiences. I adopted a reflexive approach to interviewing and analysis. Through thematic analysis and a very

iterative approach to considering the data, major themes and sub themes linked together by a meta-theme were developed. These themes were generated from the synthesis of data and aimed to reflect key issues for the young people and a deeper understanding of them.

## CHAPTER IV: FINDINGS

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### Introduction

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This chapter has been divided into four sections corresponding to themes to provide structure and clarity in presentation; these themes were generated through the analysis of the data. The aim of the chapter is to represent the journey of the young people through their descriptions of their experiences from the emergence of symptoms to their diagnosis. It is simply their story, and emphasis is placed upon stories that the young people shared; they are central to the findings. I am a children's nurse and also a researcher in a tertiary referral centre for rheumatology, therefore, I am mindful of my own participation in this process and how my own assumptions, actions and pre-conceived ideas could have shaped the data. I will address any such influences and limitations throughout the chapter using a reflexive narrative.

The four themes ('Emerging Illness', 'Seeking Help', 'Diagnosis of Lupus' and 'Resilience, Reflection and Recovery') are linked together by a meta-theme – 'passing of time' that runs through all of them. 'Passing of Time' is not a static concept; it evolves and changes and this is evident in the expression of this meta-theme within the four themes.

Within this chapter, I first present an overview of each theme before a more detailed presentation of each theme is given later in the chapter. Within each theme the key elements are presented and, as appropriate, anonymised quotations are used to illustrate and provide supporting evidence. See Table V for the demographic details and code identifiers of the young people who participated in the study. Limited information is provided about each individual young person to

reduce the risk of breaching anonymity. Disease duration refers to the time since diagnosis of lupus at the point of interview.

**Table V: Demographic Information & Code Identifiers of Young People**

Code Identifier	Time from symptom onset to diagnosis	Age at Diagnosis	Educational Stage	Disease Duration Since Diagnosis
YP1	4 years	15	Secondary Y10	24 months
YP2	3 months	11	Primary Y6	6 months
YP3	1 year	16	Secondary Y11	7 months
YP4	4 months	13	Secondary Y9	18 months
YP5	10 months	15	Secondary Y10	18 months
YP6	5 months	11	Primary Y6	28 months
YP7	3 months	16	Sixth Form Y12	29 months
YP8	4 months	12	Secondary Y8	9 months

## Overview of key themes

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### Emerging Illness

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‘Emerging illness’ encompasses the initial part of the journey to diagnosis. It is representative of the period of time when a young person first starts to notice that something is not quite right. There is a comparative element to this process and acknowledgement by the young person that they are experiencing something different to their peers. Within this theme I present the collateral effects of these symptoms and the emerging disruption to the lives of the young people. The meta-theme ‘Passing of Time’ reflects the time before help is sought to alleviate or challenge the emerging illness.

### Seeking Help

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‘Seeking help’ evolved from the descriptions of first contacts with health care services as a young person makes decisions to acknowledge their first feelings of being unwell. It encompasses how the young people (with their parents) seek to establish avenues to help them cope with and to unravel the gradual emergence of illness. Many of their reported experiences are seated in dissatisfaction and uncertainty. Whilst ‘seeking help’ a young person often begins to experience social isolation as their school attendance starts to wain and interaction with peers becomes more difficult to maintain. ‘Passing of Time’ is evident within the context of a complex and insidious onset of illness. At this point diagnosis may represent a significant challenge for even the most experienced of professionals to recognise as lupus.

### Diagnosis of Lupus

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‘Diagnosis of Lupus’ is a theme that is greater than the few words by which it is represented and is for the most part the pinnacle of the journey. It is representative of a single moment in time when a young person is presented with a name and explanation for their symptoms. A colourful

fabric of interwoven concepts is evident in the accounts of the young people. The young people broadly talk of 'Diagnosis of Lupus' as being positive with processes such as adaptation, control, independence and resilience resonating through their reflections. The journey at diagnosis is unique and dependent on the experiences, environment, political and social structures particular to each young person. Lupus, as a diagnosis, creates a degree of uniformity and similarity in this group of young people. However, Lupus as a disease can be heterogeneous in presentation and similarly the experiences of diagnosis for a young person are also profoundly varied. 'Passing of Time' is perhaps less evident in 'diagnosis of Lupus' as diagnosis is in essence representative of a single moment in time.

### Resilience, Reflection and Recovery

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'Resilience, Reflection and Recovery' was born out of young people revealing experiences relating to a point in time beyond diagnosis which had originally been the intended endpoint of the initial research inquiry. The young people's descriptions of their journeys to diagnosis were detailed and were contextualised by their reflections looking back to diagnosis. Their reflections enrich, develop and provide cohesiveness to the experiences they shared. The inclusion of 'resilience, reflection and recovery' as a theme, adds integrity and completeness to their experiences of diagnosis. It also provides a link to their contemporary and future experiences of Lupus. 'Passing of Time', is evident in the on-going nature of this lifelong illness.

## Section One - Emerging Illness

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### Introduction

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The first descriptions of a change in the health of the young people as an emerging illness are revealed in this section and are characterised by expressions of physical symptoms and the impact of such symptoms on the life of the young person.

### Emerging Physical Symptoms

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The main physical symptoms articulated by the young people were tiredness, feelings of pain such as joint pain, debilitating headaches, rash, sickness and other non-specific symptoms. These symptoms are now presented in more detail.

Tiredness was mentioned by every young person in their descriptions of first feeling unwell and it was the key 'emerging physical symptom'. It encompassed increased time spent sleeping but also feelings of fatigue due to restful sleep being compromised and interrupted because of pain. Tiredness was indicative of first feelings of being unwell, as YP 3 explained:

*'I was getting tired easier, yes that was probably it, getting tired easier and little things that I just don't know.'* (YP3)

Being tired, as an 'emerging physical symptom', was inextricably linked to the onset of pain in seven of the young people. Pain and in particular joint pain was a significant and disabling factor experienced by several young people and the second most mentioned 'emerging physical symptom'. The widespread nature of pain was also evident with YP8 explaining *'I think it was almost all of them (joints)'* and as *'really bad pains in my body'* (YP6). A gradual onset of disease and an increasing severity of pain are revealed through descriptions of painful joints, as one young person explained:

*'It just got a bit worse and worse and worse but I didn't really think about it and then it was the big joints, my knees hips and shoulders those are the first ones*

*then gradually oh and then my ankles. Hips knees and ankles they were quite bad and then afterwards it was my wrists and my elbows as well...'* (YP8)

A significant symptom in YP6 appeared to be the experience of headaches which were continuous and a source of increasing pain. As an 'emerging physical symptom', headaches were both painful and disabling as she was also sensitive to light and noise. The physical symptoms in her descriptions of 'emerging illness' are pain, tiredness and headache. YP6 recounted spending significant amounts of time in bed and appears overwhelmed by the onset of emerging illness:

*'I didn't know anyone. I just felt like I wanted to go to sleep all the time. I fell asleep during the day and I fell asleep during the night. I used to sleep twenty four/seven and I never used to like eating..... my chest used to hurt and I never used to get out of bed.... I had like really bad headaches and I couldn't have the light on* (YP6)

A strong emphasis in her descriptions is placed upon the 'passing of time' with one day merging into another as she struggles with symptoms before seeking help.

YP1 also talked about a lengthy time from first feelings of being unwell to diagnosis. The 'passing of time' theme was lengthy due to him receiving a first diagnosis of another condition and then after another period of time a diagnosis of Lupus was made, as he explained: 'It was about four years, which is quite a long time.' The 'emerging physical symptoms' for YP1 were non-specific and complicated by his first diagnosis reflecting the complex presentation of lupus in young people. YP1 reflected that his experience of emerging physical symptoms had been unusual, '*A lot was said about how it was atypical (as a presentation of lupus)*' (YP1).

In comparison, the classic facial butterfly rash and skin photosensitivity which is characteristic of lupus was recognised by only one young person as a first feature of being unwell, it was described as red, bumpy and sore on her face and chest. This symptom is visible and is unique

due to its visibility as an emerging physical symptom. The visibility of the rash evidently brought its own anxieties for her as a teenager as she recalled: *'I really don't know how I went to school with it all over my face'* (YP5). Despite such apparent visible evidence of disease YP5 was required to endure a lengthy time line from first appearance of the rash until diagnosis of lupus. Despite a timely diagnosis being crucial to successful treatment of lupus, the existence of a lupus rash in this young person did not expedite this outcome. It was nine months from appearance of the rash until time of diagnosis at the tertiary referral centre.

'Passing of time' was evident in the reports of all the young people who spoke of varying lengths from three months to four years they were in the emerging illness phase. YP3 found it difficult to be precise and explained *'I would say about eight months but I don't really know'*. On further questioning YP3 provided details of an even longer period of 'emerging physical symptoms' when she revealed that she had Raynaud's for two years that had been worsening:

*'Well I don't know because at first we just thought I must just get cold easier we didn't think anything of it until it started getting worse that was like year ten and eleven so that is about two years, when we first started thinking that it was something'. (YP3)*

It is apparent that the onset of the 'emerging illness' was difficult for the young people to define, for many it seemed the onset of physical symptoms was vague. Upon giving a retrospective account YP8's (who was diagnosed in the autumn) timeline appears to be lengthening as she continues to recall the details of 'emerging illness':

*'I think it was just like gradually and I think it was probably maybe a month before I think. I can't really remember because it was that long ago but I think it was like in the summer'. (YP8)*

YP8's description of her symptoms as having a gradual onset reflects lupus as an evolving disease process. However she acknowledged the inherent difficulty in recapturing accurately the events of that time.

It is difficult to surmise how much my own position as a research nurse within the rheumatology team at the tertiary centre influenced the mode of answers. Although the young people described a range of symptoms the language used revealed minor leanings towards medical terminology and I perceived this as synonymous with their growing knowledge about their illness rather than phrased in such a way because they were talking to a researcher who was also known to them as a health professional.

### Emerging Disruption to Life

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'Emerging disruption to life' highlights how the young people's lives gradually began to become affected by the onset of physical symptoms characteristic of lupus. Pain was discussed in relation to the activities that it prevented the young person fulfilling, things as simple as being able to brush their teeth, walk to school, and get in and out of bed. YP7 commented:

*'I could not brush my teeth properly and my Oh it was my legs and my knees they were sore. Also getting in and out of bed it was really hurting'. (YP7)*

Painful joints resulted in one young person 'walking like an old woman' (YP2). YP8 talked of struggling keeping up with peers walking to school but persisted showing remarkable determination and resolve. It is perhaps an indication of how teenagers do not expect there to be anything 'wrong' with them, especially if signs are subtle. Even when there are apparently obvious signs, YP8 did not necessarily associate them with anything untoward even when she is able to make a comparison between herself and her peers. However, there comes a point when she realised that she was not able to do the same things as her friends, YP8 explained:

*'It got a bit harder walking to school and it was just a bit more harder and I just found it more difficult to keep up with everyone else walking and that just got a bit worse and worse and worse but I didn't really think about it..... I just carried on walking to school as I didn't really think it was anything'. (YP8)*

It is apparent from the above that she was able to detect that her ability to walk to school was deteriorating. However, it is difficult to determine whether the young person delayed seeking help through either a lack of awareness of her own health needs, or a refusal to admit that something was wrong. YP8 could have been going through a process of internal realisation/adaption towards her altered health; a process necessary for her to move onto the next phase in seeking help to address her health needs. In contrast, eventual diagnosis with lupus was not a surprise to YP7 who had realised that there was a significant problem with her health as her ability to function had deteriorated:

*'I knew there was something wrong with me because I could not do anything'.  
(YP7)*

The way school life was disrupted for the young people differed as did the emerging illness. YP4 was unable to attend for many months prior to her diagnosis, whereas YP3 was able to attend and complete her GCSE examinations despite repeated outpatient hospital appointments during this critical period of her education. YP4 detailed a period of two months when she was at home unable to go to school prior to being assessed by a doctor. Although YP2 had attempted to go into school she had felt unwell and had to come home; she was absent from school for five weeks before diagnosis. YP5 who suffered a visible lupus rash was required to stay away from school as a GP thought it might be infectious. Repeating cycles of illness and absence from school, are evident in the description given by YP6 who said:

*'I would be better for about a week then I would go back to school and then on the weekend I would be ill again and I will go to school for three weeks then it would just keep going like that...'* (YP6)

She talked of great disruption to her life due to the emerging illness. Such were the severity of her symptoms she withdrew from family life and normal functioning because of all encompassing feelings of being unwell. She explained:

*'I used to tell my mum to leave me in the dark and I'd switch off the light. I'd close all the curtains and hide under the duvet and just fall asleep and I didn't like the TV on. My little brother was upstairs making a noise and I didn't like that and I just liked to be alone and quiet and I just felt really bad in myself and I just wanted to sleep and not talk to anyone.'* (YP6)

What is apparent is that all the young people experienced some disruption due to the emerging illness. For all of the young people the timing of onset of physical symptoms and disruption to life occurred at a stage in their education that was significant for future foundations of learning and direction such as during academic examinations or transition from primary to secondary school. Furthermore for each young person the emergence of illness was both a significant and lengthy period of time that differentiated them from their peer group at a stage in their development when good health was both expected and desired.

## Section Two – Seeking Help

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### Introduction

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This explores the experiences of the young people when their health concerns have escalated to the point they seek help from a health professional. Although in all cases first contact with a health professional was with a General Practitioner (GP) each young person had a different experience of seeking help.

### Experiences of Dissatisfaction

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‘Experiences of dissatisfaction’ with the help they received surfaced in all the young people’s stories. As they struggled with their emerging illness a point was reached when either they or a family member made a decision that further help was needed to bring about a change to the current situation. YP2’s mother commented on the tipping point that initiated their ‘experiences of seeking help’; this was closely followed by ‘experiences of dissatisfaction’:

*‘I think the final straw came when I had to try and carry her up the stairs to the toilet. And I thought this just isn’t right’. (YP2 mother)*

YP2’s mother had been asked to wait for a set period of time before returning for another appointment. When ‘tipping point’ was reached she was dissatisfied with the GP’s advice and therefore decided to bypass primary care services and seek help at the tertiary care centre.

It is difficult from retrospective accounts to wholly untangle ‘experiences of dissatisfaction’. However several of the young people recalled how they repeatedly visited the GP and YP2’s mother talked about how the GP did not seem to believe them when they described her symptoms:

*‘So we went back and he was basically asking me because she had time off school it seemed the GP didn’t believe me.... He was asking questions like; ‘Is there any reason she doesn’t want to go to school’? And that’s not something, that’s not*

*and I wouldn't bring her if I didn't think something was wrong, so that was a bit off putting' (YP2)*

YP7 sought advice after dissatisfaction with her own GP from a family friend who was also a doctor. Armed with advice from their family friend her father insisted on blood tests being taken. Whilst YP5 and family escalated their concerns and requested a private referral due to dissatisfaction with their family doctor:

*'Mum was like 'We're not getting anywhere' (with the GP) so we went to the private hospital'. (YP5)*

In retrospect the young people have the benefit of hindsight and they are now aware that the initial symptoms that triggered the visit to the GP were because they had lupus. Their dissatisfaction seemed to stem from their expectation that a doctor should be able to recognise and diagnose any medical condition, particularly one such as lupus, which is a life-long condition that can have enormous consequences for a person's life. As a children's nurse I am well aware of the multi-factorial limitations and difficulties placed upon health professionals. Time at a GP appointment is always limited and a history with examination performed under this time pressure is only a snapshot. The responsibility of both young person and family to convey detailed personal symptom information is challenging and the GP is required to respond accordingly. However, again, the diagnosis of lupus has to be reconciled with a gradual onset of the symptoms and the insidious nature of disease. It would also be worthy of note to explore and contrast the experiences of GP's who are the first contact for these young people and are also unlikely to have seen a case of lupus in childhood. Unfortunately this is beyond the scope of enquiry at this time.

The expectation that a GP is able to recognise and diagnose the features of lupus is apparent in the comments of YP4 who after repeated visits saw a different doctor who initiated the referral to the tertiary centre. She explained:

*'We went to see a better doctor. They kept on asking me questions and I answered them. They thought I should be admitted straight away'. (YP4)*

It is clear from her description that YP4 had finally been offered an intervention that had the necessary urgency and meaning. At this point she has been 'poorly' for several months and her condition was rapidly worsening, it is difficult to imagine how YP4 must have been feeling. What is striking is there is absolutely no doubt that the GP acted appropriately at that time. YP4 elevated this doctor to '*better*' status, the help she had desperately sought had finally been realised and the reason for her feelings of being unwell began to be explored.

### Feelings of Uncertainty

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'Feelings of uncertainty' are closely linked to dissatisfaction, but are also a distinct characteristic as they are associated with the feelings of the young people and their families during and prior to their initial contacts with the GP, at a time when they are questioning/deciding whether to seek help and also which route to take in doing so. The young people's realisation that something may be wrong was associated with feeling uncertain as to the appropriate avenue to convey their concerns. The parent of YP2 sought help and when that was not offered they experienced dissatisfaction towards their GP; this in turn led to them experiencing uncertainty and consequently initiated their own heightened response. For this particular parent the decision to self-present at the emergency department of the tertiary referral centre was taken, rather than make another visit to the GP.

Other experiences of uncertainty are related to the initial diagnoses suggested by the GP and in particular the mention of a 'virus' as a possible diagnosis and of 'paracetamol' as a treatment. Both are evident in several of the young person's accounts. YP4 sought help at a walk-in centre in the early hours of the morning after feeling particularly poorly. She and her father presented outside

of conventional hours providing an indication of their perception of the severity of her symptoms; this suggests YP4 felt she was nearing a crisis point. At the time of 'seeking help' YP4's mother was heavily pregnant with her fifth child. It is apparent from her account that she felt there was uncertainty in both diagnosis and treatment. Interestingly the GP and the walk-in centre proffered the same advice:

*'We went to the walk in. It was about two in the morning or something. They said they think it was a virus and gave me paracetamol. I remember taking the tablets and everything but nothing it never worked. (Went back to GP) They said the same thing they think it's a virus and gave me paracetamol'. (YP4)*

'Feelings of uncertainty' also led to YP6 questioning whether to seek help again after repeated presentations at GP and hospital. When her symptoms escalated she displayed a lack of faith in the hospital to provide the help she needed due to repeated dissatisfaction in the preceding months:

*'I felt like I couldn't breathe properly and I said to my mum like I can't feel my heart properly and my mum said you are going to hospital and I said that I didn't want to go to hospital because I knew they wouldn't do anything because I've been to hospital before...'* (YP6)

'Feelings of uncertainty' also related to how the young people and families rationalised seeking help. YP3 talked about visiting the GP to check if she should be experiencing certain symptoms *'just to see if that was right or not'* (YP3).

For several of the young people 'feelings of uncertainty' reflected the insidious creep of lupus presentation in young people. Even the young people were sometimes unsure about their own feelings of being unwell until, as illustrated by YP6 and YP4, a crisis point is reached when symptoms are so severe that emergency inpatient treatment is needed. Once again

'passing of time' is evident throughout during repeated presentations at both primary and secondary care, whilst symptoms escalate, initial treatments fail and diagnoses are suggested.

## Section Three - Impact of Diagnosis of Lupus

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### Introduction

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For all of the young people this single point in time was significant as they all experienced a change in health status from a young person without lupus to a young person with lupus. The young people talked about how diagnosis impacted on them but also how their family was affected and the exploration of these interlinked themes guides the narrative.

### Impact on self

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All of the young people were unaware of lupus as a disease prior to their diagnosis. One young person had some recognition of the word lupus and recalled the association with the butterfly due to a building close to her primary school displaying the symbol. For several of the young people this lack of knowledge may have protected them from feeling distressed and reduced any negative 'impact on self' at the time of diagnosis. YP7's response was fairly typical:

*'I had no idea what it was so I did not say anything and it did not scare me that much. I did not think a lot about it'. (YP7)*

It is apparent that YP7's perceived her lack of knowledge as protective; it reduced her ability to make assumptions or judgments about her future health status as a young person living with lupus. Another young person also talked about her lack of knowledge of lupus. However after asking more questions she became frightened by the severity and upset by the specialised lupus diagnosis:

*'Well not so much because I didn't know what lupus was I just thought it was like a normal sort of thing because I'd heard of diabetes and stuff but I never heard of lupus but then when I asked my mum what is was she said my body doesn't make*

*enough white blood cells and I thought that it must be something serious and I got a bit upset, scared'. (YP6)*

YP3 supported the view that a lack of knowledge could be positive because she was able to respond to her diagnosis without the burden of preconceived misconceptions about lupus. It allowed her to be educated about her disease from a team experienced in delivering specialised care and minimised the opportunities for inaccurate information giving as she explained:

*'I didn't really think anything of it because I just had never heard of it, whereas if it was something else like you had heard of, then that's like 'Oh God' but then (Rheumatology Consultant and Lupus Nurse) started talking about it a bit like not worrying but to find out what it was and stuff'. (YP3)*

YP4 was in hospital for a month before a conclusive diagnosis was made – she uses the word 'noticed' - and this perhaps reduced the 'impact on self' of the eventual lupus diagnosis:

*'They kept on saying she might have this, she might have that and then in the end they noticed that I had lupus'. (YP4)*

However, during the period of not knowing she was suffering from lupus, YP4's experienced heightened anxiety, she commented about this period saying:

*'It was like 'Oh my God am I going to die or something''? (YP4)*

For YP4 'impact on self' of obtaining her diagnosis of lupus was a sense of relief in contrast to the 'emerging illness' and 'seeking help' phase. She commented on her diagnosis: *'thank god they know what it is'*. A sense of relief about diagnosis was also evident in YP3's account:

*'It was more a like a sense of relief as well knowing what it was and that you could be treated rather than wondering what it was'. (YP3)*

In comparison (YP8) found it difficult to recall the moment of diagnosis. She had experienced an extended period in hospital whilst a diagnosis was made. YP8's account seemed passive in terms of

her involvement with the process of diagnosis; she seemed almost removed from what was happening around her as she explained:

*'Not really I think they did scans and things then they realised it wasn't (arthritis) because I think I started getting symptoms that weren't just the arthritis. I don't really remember when what, they did lots of tests and they realised it wasn't that but I can't really remember'. (YP8)*

She appeared unready to learn about lupus and it is difficult to surmise if this was an intentional passing of responsibility onto her parent or a protective mechanism to prevent any confrontation with her own mortality. Her perceived 'impact on self' is minimal but I am left wondering about her repetitive use of the word 'really' and whether this is an indication that YP8 is experiencing denial about her feelings and not yet ready to admit that she has been deeply affected by her diagnosis:

*'I didn't really want to (look at information on Lupus) my mum did. I think they sent home like and booklets and my mum looked at those but I didn't and I still don't now. I don't really because I don't really worry about it'. (YP8)*

Previous experience of hospital and illness resulted in an altered impact on self for YP1. He felt that having been diagnosed with another disease prior to lupus lessened the impact of his lupus diagnosis but did not prevent him articulating awareness of the severity/consequences associated with diagnosis when talking about his experiences:

*'I think it was quite scary at first, especially with the (first) disease and I thought I have got this for my whole life, but I think with the Lupus, it wasn't as bad because I already had the (first) and so once you have already got something it does not hit you as much. With lupus I have not had many problems at the minute, but it is quite frightening as it can be serious'. (YP1)*

However he also commented that lack of awareness and understanding of lupus contributed to him feeling worried:

*'I think because it was all very foreign and lots of blood tests and I did not know what it could be at all and that can be is quite worrying sometimes'. (YP1)*

Resonant in his descriptions of diagnosis are the uncertainties surrounding identifying lupus as the cause of his symptoms. Even at diagnosis he remembered being told he had three rather than four of the ACR criteria needed for classification of lupus. He was also able to acknowledge the inherent difficulty for clinicians in making a diagnosis:

*'I think it would have been better if you were more certain, not a guess, but symptoms is not a lot we can do about'. (YP1)*

YP2 reacted differently to her diagnosis as she did not understand it. Her first priority was to obtain discharge from hospital to the security of her home environment. She equally found all the investigations daunting. Interestingly in contrast to the experiences of other children (YP1, YP4, YP6 and YP8), YP2's mother recalled the term lupus being associated with her daughter's illness within a short time of self-presenting at accident and emergency in the tertiary referral centre:

*'Do you know as soon as we came to A and E, I remember one of the doctors writing Lupus with a question mark as soon as we came in because she had the flush on her face and the first doctor said it could be lupus'. (YP2 Parent)*

For YP2 the 'impact on self' of a swift diagnosis had been overwhelming. It was clear from her story that it had been both unexpected and traumatic, yet medically it was a perfect lupus diagnosis happening within twenty-four hours of self-presentation.

The 'impact on self' of diagnosis was dependent on many variables including context or place of diagnosis. YP5 was the only young person offered a diagnosis outside the tertiary centre. The dermatologist at a private hospital suggested lupus as a possible diagnosis prior to her appointment at the tertiary centre and she explained:

*'Yes straight away pretty much I think he knew what it was yes'.(YP5)*

Of all the young people YP5 approached her account pragmatically and detailed her journey to diagnosis in a simple step wise statement:

*'You think; I have nothing wrong and then I had this rash and then I have lupus so.....'.*(YP5)

It is obvious that the 'impact on self' is subject to many complex processes and the journey at diagnosis is unique dependent on experiences, environment, and social structures of the young person. As health professionals we are always searching to deliver the best possible care for children and young people. As a children's nurse, listening to the accounts of the young people, the need for both an individualised and specialised approach to diagnosis resonated loudly. At the heart of all the diagnoses are young people who deserve care of the highest standard that is delivered in a timely and appropriate manner according to their individual needs.

### Impact on family

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All the young people when given their lupus diagnosis had family members present during that single moment in time. However, the 'impact on family' of diagnosis also acknowledged the effect on the family before diagnosis and as a result diagnosis. 'Passing of time' is also evidenced as the family unit had adapted to a change in circumstances. In some families fragmentation occurs because a parent and the young person are isolated away from the family unit during long periods of time spent in hospital. In YP1's family, family bonds are reinforced as YP1 spends greater amounts of time in the company of both parents and is able to talk to them about his illness. Another young person talked about separation from her sibling and the difficulties encountered by her well sibling due to worry and concern over her diagnosis and treatment (YP2):

*'She was very upset because when I was at school she would sometimes play with me and my friends so when I wasn't there she didn't like talking to my friends...*

*she couldn't stop being reminded of it because everyone kept coming up to her and asking why I was off'. (YP2)*

Things had become so difficult for the younger, well sibling that YP2's parent admitted she had allowed her to take time off school in an effort to keep the family unit together, taking measures to strengthen family bonds during the crisis. An older sibling had also shouldered the burden of care for the younger sibling during this time and YP2's parent recognised that medical interventions had effected family functioning and made efforts to normalise family dynamics.

Further evidence of the fragmentation of family is evidenced in the account of YP6. She talked about the wide reaching impact of her diagnosis on the family unit. Her mother struggled with guilt due to increased time spent away from her younger child versus time spent in hospital at the bedside of her very sick elder child:

*'He [younger brother] cried a lot and he didn't feel he was getting cared for. My mum tells me now that she was so taken in by my illness that she forgot that she had a son and another daughter because she was always there and she felt really bad afterwards because she couldn't look after my brother as much as she looks after me and my sister when we were young..... my sister had to look after my brother all the time'. (YP6)*

YP6 appears insightful, showing maturity and awareness surrounding the impact of her diagnosis upon the whole family and the burden of care that her sibling picked up:

*'She [elder sibling] was in year 9 and she wanted to go out with her friends and she wanted to have fun but she had to cook for my brother, she had to do like make his bed, get him dressed, give him a bath, do everything for him and do everything for herself as well so she never had time for herself'. (YP6)*

Other young people revealed separation from their families as parents tried to divide their time between being with them in the hospital during daytime hours and returning home to care for

their well siblings overnight. Hospitalisation is significant in many of the young people's accounts and its 'impact on family' should not be underestimated. It is clear that several of the young people were aware that their illness was not just personal to them but a process that was multi-factorial and had an 'impact on family'.

*'I think my mum just worries all the time but I think she's just like that and then she goes and reads on the internet about other people and I don't like that she gets worried'. (YP8)*

Worthy of note are the views of YP5 who talked about the effects of her diagnosis on her family. She framed diagnosis as a necessary conclusion rather than a starting point for a lifelong journey of disease management:

*'I think my mum was basically glad that it was sorted out, she was just glad it was sorted'. (YP5)*

This is perhaps a result of her pragmatic approach to diagnosis, her positive approach to treatment or maybe a reluctance to fully engage with her health status as a young person with lupus. She commented:

*'I kind of thought I might grow out of it and see if it passes... Hopefully yes. Carry on with my medication and see what happens'. (YP5)*

This type of reaction toward diagnosis provided a springboard to the next theme as all the young people talked about their contemporary experiences and their future.

## Section Four - Resilience, Reflection and Recovery

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### Introduction

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This section warranted inclusion as it contextualised the experiences of the young people throughout their journey to diagnosis. 'Resilience, reflection and recovery' are characterised by things that went well and things that did not and placed in context by the young people as they looked back on their experiences of diagnosis. 'Passing of time' relates to both time since diagnosis and the future for the young people with lupus.

### Resilience

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Several of the young people displayed evidence of 'resilience' despite their diagnosis, they were able to adapt given the knowledge, support and tools to do so. Adaptability was illustrated by many young people drawing on coping skills that appeared to be supported by a youthful 'resilience' towards adversity. YP3 commented on how she had adapted to taking her medication:

*'At first I struggled to get into the routine but now it is just a part of everything'.*

(YP3)

Taking daily medication is integral to successful disease management and almost all of the young people had not experienced routine tablet taking before. YP4 emphatically detailed her difficulty swallowing tablets although explaining that she eventually adapted to the regime as she began to experience normalisation in her health status and adherence helped her to manage her disease. YP2 commented on her own rapid introduction to medicines:

*'I got used to them after a bit but then I didn't like the medicine and I couldn't take the tablets, I took one and it didn't have a coating on and it put me off. It was paracetamol in A and E and it put me off. So it was daunting thinking you had to take all those medicines'.* (YP2)

It was her parent who added perspective and illustrated the ‘resilience’ of YP2 in taking her medication to promote recovery despite feeling challenged by the process:

*‘I think she would have taken anything to make her feel better because she was so unwell’.* (YP2 parent)

A youthful ‘resilience’ was also apparent in the way YP8 was able to minimise the impact of her diagnosis by normalising it and emphasising recovery rather than disease as key from her perspective:

*‘I never really thought it was a condition. I never really react to it like a condition. I never really thought it was serious or anything which it obviously can be but I never really thought of it like that so it wasn’t a big thing no. It was good they found out they could treat it... .. I didn’t really think of it as a bad thing that I had, I just wanted to get better’.* (YP8)

Normalisation and ‘resilience’ of the young people towards their diagnosis appeared crucial to them in the achievement of an optimum health outcome. However although YP6 considered the maintenance of health as her goal, she also realised that lupus would be part of her life forever and that lupus differentiated her from others whose reality was to live without a life threatening illness. She explained how she dealt with lupus:

*‘Just pretend like you’re normal and like everyone else, just forget about the medicines and everything, just lead a happy life with your family and your sisters and brothers, don’t let anything get you down, be normal and happy and just be like that you’ve got no illness, just be like everybody else’.* (YP6)

## Reflection

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Talking about their experiences from first symptoms to diagnosis allowed the young people the time to reflect upon their journey from a young person to a young person with lupus. I felt

privileged to be able to listen to their stories and all the young people (with the exception of one) did this without the presence of a parent. Diagnosis was acknowledged as a crucial point for YP5, she realised that the identification of lupus was pivotal to her experience but her lengthy time to diagnosis she identified as negative. Other young people had similar negative reflections about the lengthy period of time taken from first symptoms to diagnosis:

*'I think I would have liked to have known what was wrong with me sooner but that perhaps is just me. I think everyone is different but I quite like knowing'. (YP1)*

YP6 reflected on her experiences of different treatments used to manage her lupus. Significantly YP6 is currently enduring maximum treatment and her maintenance of health is subject to regular inpatient treatment regimes. She talked of the restrictions placed upon her by medications and revealed the psychological impact her illness was having:

*'If I was just on the medicines twenty-four seven and I just had to sit around watching TV I'm not moving away from the bed and I did and I just didn't feel like me because I am an active person. I like going to parks and I like going to my friend's house. I like playing outside in the garden and stuff and it just felt like I was trapped in a cage and I wanted freedom and I just didn't feel like it was me'. (YP6)*

When the young people talked about the restrictions placed upon them as a result of their diagnosis, school featured highly in their reflections. Some aspects of their disease created what they felt were unwarranted attentions (YP1) and this was challenging for them to deal with. YP8 recalled the problems encountered educating school about lupus whilst the family themselves were still learning about the complexities of the disease. She commented:

*'I think the teachers they didn't really understand, I think they thought they understood. I don't think they thought they will be nasty about it but I don't think they understood and then and then they maybe do something and then I couldn't*

*do it. It made me feel really uncomfortable they didn't understand and I think that was probably quite hard'. (YP8)*

Social interaction with peers was also affected, maintaining contact with friends was important and many of the young people talked about the social isolation of their illness:

*'It sort of affected me when I used to hang out with my friends and that sort of thing declined and the combination of a) to do my school work but b) I did not have the energy to do it'. (YP1)*

For some young people communication between school and home had deteriorated as a result of continued absence. This added extra pressure with increased worry about academic attainment. A positive outcome during academic examinations was achievable and although YP7 did think that her grades were going to be affected, she attained good results and successfully commenced university.

## Recovery

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'Recovery' provided a context from where the young people could talk about the onset of symptoms and experiences leading up to and including diagnosis. For all of the young people 'recovery' was associated with things going well and improvement in their health status. YP7 contextualised the difference:

*'Yes there is a big difference. Yes now I can do most things, again...But before I could not brush my teeth properly or get into the car'. (YP7)*

Several of the young people did not perceive any restriction placed upon future aspirations due to having lupus. Other young people had not considered how their altered health status might impact in the long term.

Treatment was associated with recovery and many of the young people sought to place emphasis on recovery as an achievable outcome. Significantly YP6 was the only one of the group who talked about a future blighted by an enduring complexity and longevity of disease:

*'I don't like to think about the future, people always ask me like 'Will lupus ever go away'? They asked me 'Will I ever stop going to hospital'? And I don't like it when they asked me that because I don't think about it myself because it upsets me. I always think that if I'm on treatment now sometimes I'll get ill, sometimes I don't get ill, sometimes medicines work, sometimes they don't'. (YP6)*

## Conclusion

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The thematic analysis generated the key themes and the meta-theme that reflect the experiences of the young people who participated in the study. The young people were able to describe their experiences of their 'journey to diagnosis. I begin to unpick and untangle their stories through detailed discussion in the next chapter.

## CHAPTER V: DISCUSSION

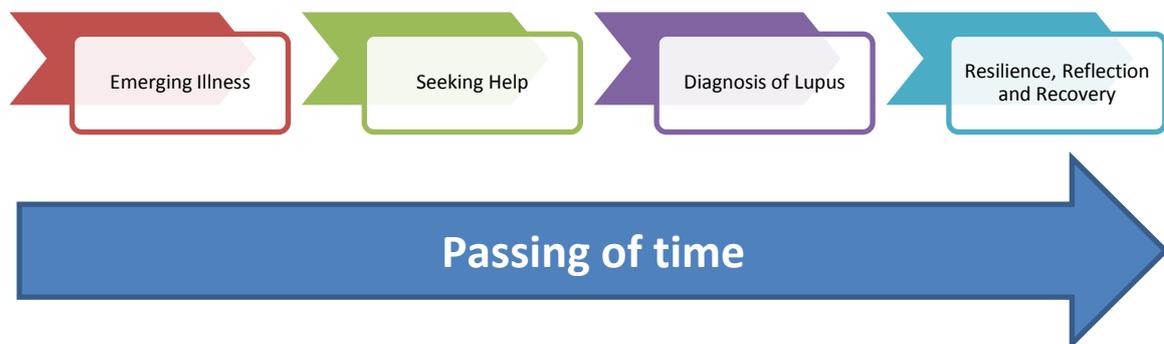
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### Introduction

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All the young people told their story of 'symptom onset to diagnosis' each describing a journey that was personal to them displaying commonality through a shared disease. Thematic analysis of the data generated four themes - 'Emerging Illness', 'Seeking Help', 'Diagnosis of Lupus' and 'Resilience' Reflection and Recovery'- that are linked together by a meta-theme 'passing of time' (the nature of the expression of the 'passing of time' and its perceived speed changes in each theme) (see Figure 5: for an illustration of the themes generated). Within this chapter I present a discussion of the themes using a reflexive narrative supported by relevant literature addressing limitations and detailing recommendations for practice.

I purposely excluded papers from the literature review relating to adult lupus. My intention through doing this was to immerse myself in the experiences of young people rather than infer meaning from adult studies as is often seen in the management of JSLE (Beresford 2009). However, where appropriate within this discussion, where there has been an absence of appropriate literature focussing specifically on young people, I have drawn links between my findings and those of authors whose work has considered adult populations. These links show interesting resonances although there are clearly limitations in trying to synthesise links between populations with different needs and at different developmental stages of their lives.



**Figure 5: Illustrating four main themes & their relationship to meta-theme**

### Emerging Illness

This theme is critical to the beginning of the young peoples' story as any narrative is grounded in some form of starting point or introduction. However for the young people this starting point or introduction was difficult to explain as they did not experience a clear onset of symptoms. Difficulties in distinguishing between normal adolescent symptoms and those of a severe disease such as lupus have been recognised (Kelly 2012). The young people found the beginning of their journey the most challenging part of their 'journey to diagnosis' to articulate and sought clarification from me about how and from which point to start their story. I am mindful that this was also the start of the in-depth interview so this may have also simply been part of settling-down behaviour. This also resonated with Woodgate (1998) who observed that adolescents found it difficult to describe their experiences of chronic illness with the exception of when questions centred around feelings, and reported the initial request of 'putting it into words' as being difficult. Once a starting point was established many of the young people reflected on symptoms they felt might have been associated with emerging illness. It was through the description of physical symptoms and the

disruption to life as a result of such symptoms that the young people were able to discuss their illness.

Tiredness for the young people was the key 'emerging physical symptom' identified. Fatigue is well documented as a presenting feature of JSLE (Hoffman et al 2009, Kone-Paut 2007, Levy & Kampius 2012, Papdimitraki & Isenberg 2009) and has been recognised by young people with lupus in other studies as a significant factor in restricting participation in activities (Houghton 2008). Interestingly, fatigue, although recognised as a symptom of JSLE, is not included in the American College of Rheumatology Classification Criteria (Hochberg 1997) for diagnosis of lupus. Emerging symptoms of lupus differ from the clinical classification criteria for diagnosis (Stockl 2007) and therefore require investigation into the cause of symptoms. For example, fatigue can be symptomatic of a low lymphocyte count or a result of inflammation in the joint (arthritis) both of which are included in the criteria used when making a diagnosis of lupus. As previously mentioned, young people require four of the criteria - although not simultaneously - for a consultant paediatric rheumatologist to make a diagnosis of lupus. Tiredness or fatigue in addition to such criteria adds weight to diagnosis and fatigue is thus recognised as a non-specific symptom rather than a clinical criterion for diagnosis. Although present in all the young people other features of lupus have to emerge before a diagnosis of lupus is considered, particularly during adolescence (Kelly et al 2012). Tiredness is an unremarkable symptom in the context of their lives and their stage of development.

The young people in this study were aged eleven to sixteen years old when they were diagnosed with lupus and this is also the time when the body is 'tasked' by the changes that occur as part of adolescence (Beresford & Davidson 2007). The period of adolescence is marked by profound biological changes which impact on emotional and social development due to the transitions which occur as part of pubertal processes (Patton & Viner 2007). It is almost expected by parents that during adolescence young people will sleep longer and later. Therefore, since there is a high prevalence of fatigue in healthy adolescents (McDonagh & Jordan 2009), the difficulties encountered

by the young people with lupus in detecting fatigue symptom related changes is not remarkable. Therefore it took time for tiredness to progress from something symptomatic of adolescence to a symptom severe and persistent enough to warrant further investigation. The adolescent period is recognised as a time of transition in terms of responsibility for self-care (Sawyer et al 2007). Thus, this time of flux between the young person gaining autonomy for self-care and the relaxation of parental scrutiny might negatively influence recognition of symptoms as neither party is taking the lead as new caring boundaries are in the process of being established. Thus the physical and other changes occurring in adolescence can create barriers to symptom recognition; developmental change and gaining autonomy are both implicated (Bury 1991).

The second most mentioned 'emerging physical symptom' was pain that was widespread, increasing in severity and also described as occurring in different joints. The young people with lupus were able to articulate their pain in rich detail contrasting with literature where young people found it difficult to describe their experiences of chronic pain (Carter et al 2002). Headache was the source of pain for one young person who provided graphic and illuminating descriptions of her experiences whilst suffering symptoms of headache, pain and fatigue; a combination which proved isolating and all encompassing. Headache, although manifesting as pain, is often considered separately in the assessment of symptoms (Collins et al 2000, Goldman et al 2006) and this was also reflected by the descriptions of the young people with lupus. Being in pain can have a profound effect on the lives of young people significantly reducing their ability to participate in activities, and particularly during adolescence this may be harmful (Castle et al 2007). Experiences of pain as an 'emerging physical symptom' in young people with lupus were pertinent, indicative of suffering and expressed as 'emerging illness' during adolescence a crucial stage of growth and development.

One young person described their experiences of emerging physical symptoms as 'atypical' with the reasoning given being that presenting symptoms were unusual, resulting in a lengthy time from

onset to diagnosis. Conversely, a young person with a facial butterfly rash and skin photosensitivity both characteristic of lupus that despite its 'visibility' also resulted in a lengthy time to diagnosis at specialist centre. In the experiences of young people with lupus, visibility of symptoms, neither facilitated a diagnosis nor expedited referral. In this study, as seen in other studies, considerable differences exist in the length of time from onset of symptoms to diagnosis experienced by young people accessing rheumatology services (Cervera et al 2009, Foster et al 2007, Shiff et al 2010, Smith et al 2013). An extended period suffering from symptoms prior to lupus diagnosis is synonymous with the experiences of adults (Hale 2006, Waldron & Brown 2007, Waldron et al 2011, Waldron 2012). The experiences and challenges of the young people in achieving a lupus diagnosis were multifactorial, and varied, as also seen in Smith et al.'s (2013) study. It is also possible to see that similarities exist between the experiences the young people reported and the adult lupus diagnostic journey as characterised as diagnostic vertigo by Price & Walker (2012).

Emerging physical symptoms were contextualised by the young people as they talked about the short-comings they experienced arising from these symptoms in the real world scenario when they talked of their experiences of 'emerging disruption to life'. As in other studies, the young people were able to describe how their symptoms were affecting their ability to achieve and participate in the activities of daily living (Woodgate 1998, Moorthy 2004). This impact is also evidenced in quality of life scores (Moorthy 2012) where high scores denoted the negative impact lupus had on the daily lives of adolescents (Sousa & Guedes 2011). This was particularly notable in adolescents with recently diagnosed lupus where low scores reported reduced physical, emotional and school functioning (Tuck et al 2012). For the young people with lupus, pain once again featured highly by causing most disruption and therein restricting participation in activities. Remarkably symptoms escalated dramatically before some of the young people acknowledged their presence, acted on them and eventually sought help by visiting their GP. As seen in young people in this study, teenagers have many health concerns but may not always tell their GP about them (McPherson

2005) and experience dissatisfaction with the care they receive from primary services and especially GPs (Davies et al 1999, Jacobson et al 2000, Jacobson et al 1996, Jones et al 1997, Kari et al 1997, McPherson et al 1996). The delay in recognising and seeking help for emerging illness was evident from the young people's accounts but the reasons for this delay was not completely clear in their accounts.

## Seeking Help

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The first contact with health services for all the young people was a GP who by occupation is a medical 'generalist' and seen as a 'gatekeeper' to the National Health Service in the United Kingdom (Simon & Riley 2013). Fulfilment of the 'gatekeeper' role by a supportive GP in signposting patients with symptoms suggestive of lupus to appropriate rheumatology services is essential (Narain et al. 2004). The young people and their families were 'seeking help' for non-specific symptoms of the sort that a GP would see very often and in the vast majority of cases would be transient self-limiting problems. There was an expectation from the young people that their GP would be able to identify and diagnose the features of lupus. Indeed, the young people's expectations were similar to those of the adults with lupus in Waldron et al.'s (2012) study who were surprised by a lack of knowledge and recognition of lupus demonstrated by GPs. In addition young people may find it difficult to communicate their health needs to their GP (McPherson 2005) and this was also experienced by some of the young people whilst seeking help for symptoms of lupus.

It was only when the young people's physical symptoms affected their functioning and became unmanageable that concerns were escalated either by themselves or their parents in self presentation at emergency care or by referral to secondary/ tertiary care initiated by their GP. This point where concerns reached a point of escalation has been described previously as a 'tipping point' (Gladwell 2000, Tinetti & Fried 2004). However, even when this tipping point has been

reached and concerns have been escalated, a diagnosis of lupus in young people is not straight forward (Beresford 2009, Watson et al 2012, Watson 2011) as illustrated by three of our young people having an extended 'passing of time' whilst 'seeking help' in tertiary care. Narain (2004) notes that even the 'experts' may struggle in making a diagnosis of lupus. The young people's experiences of dissatisfaction were deeply rooted in 'seeking help' for symptoms that were not acknowledged or respected as being potentially serious. These feelings of dissatisfaction were further compounded by the realisation that even the experts may lack the answers to provide adequate explanations and are uncertain as to the reason for their symptoms.

'Feelings of uncertainty' were provoked in the young people through four types of experience: questioning whether to seek further help and initiate heightened response after dissatisfaction with their GP; in response to initial suggested diagnoses and treatments; after repeated presentations at both primary and secondary care; and experiencing an insidious creep of lupus related symptoms. Adult studies have centred upon the impact of diagnostic uncertainty during the period between onset of symptoms and diagnosis (Waldron & Brown 2007). Literature reflecting the experiences of adults seeking a diagnosis of lupus has also identified experiences of dissatisfaction conveyed through diagnostic uncertainty (Hale et al 2006, Price & Walker 2014, Waldron et al 2012, Waldron et al 2011, Waldron & Brown 2007). Although similarities appear to exist between both the young people's and adult's 'journeys to diagnosis', subtle differences are also present between the two groups. Young people's 'experiences of dissatisfaction' were associated with 'seeking help' for symptoms rather than pursuing a diagnosis and 'feelings of uncertainty' were associated with not knowing where to turn after seeking help had resulted in an unsatisfactory outcome and symptoms persisted. Therefore young people experienced dissatisfaction and felt uncertain whilst seeking help and this was distinct from but closely associated with their diagnosis of lupus. In comparison, the adults' 'uncertainty' was synonymous with both their diagnostic and misdiagnosis experiences

(Corbin & Strauss 1985, Nettleton 2006, Price & Walker 2014, Waldron et al 2012, Waldron et al 2011, Waldron & Brown 2007).

## Diagnosis of Lupus

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The diagnosis of lupus was a significant time point for all of the young people as they experienced a change in health status from a 'healthy young person' to a 'young person with lupus' and parallels can be drawn with Kralik et al.'s (2000) qualitative study of women's experiences of 'being diagnosed' with chronic illness who reported similar changes. This transition between 'being healthy' and being a young person with a chronic condition challenged their sense of who they were. As Kaplan and Baron-Epel (2002) discussed, health is a concept that is deeply rooted in individual perception, judgment and social circumstance. Although no lupus studies could be found that drew on young people's experiences, the findings from Goodman et al. (2005) demonstrate that adults with lupus perceived major physical, social and psychological consequences resulting from their diagnosis. Some of the young people felt relief whilst others were worried by the implications of their condition, concurring with research findings in adult lupus patients (Hale 2005, Waldron 2012). For each young person their diagnosis had affected both themselves and their family as seen in the findings from other studies (Eiser & Berrenberg 1995, Seiffge-Krenke 1998, Varni & Wallander 1998).

All the young people disclosed that they were unaware of lupus as a disease and lacked any prior knowledge about lupus being a condition that could affect young people. This is consistent with findings in adults (Giffords 2003, Goodman 2005, Waldron et al 2011) and hardly surprising as lupus is even less notable as a disease in young people. Many young people perceived diagnosis as an opportunity to learn about lupus as they were provided with recognised sources of information. They did not view their lack of knowledge negatively and they appeared to approach the unknown (lupus) without fear. Whilst some young people wanted to read and obtain knowledge

independently about lupus, others did not and preferred this information gathering to be done by a parent. Detailed information supplied at diagnosis prevented searching or information gathering from unreliable or unknown resources; Waldron (2011) notes this can lead to heightened anxiety and increased fear. All the young people at diagnosis received detailed written and verbal information about lupus in young people – this is probably a reflection that this study was a single centre study. However, adults in Waldron's (2011) multi-centre study were subject to different approaches, delivered by different teams. The question arises as to whether the differences in adult's and young people's experiences at diagnosis simply reflect the differences in how the diagnosis is delivered by the healthcare team? Findings suggest that a complex interplay of factors exist that have to be negotiated by young people during their 'journey to diagnosis' these are not dis-similar to the adult experience and 'nuances' in care delivery can improve or impair experience.

The young people in this study responded to their diagnosis of lupus in different ways. Other chronically ill young people have been shown to determine meaning through their experiences of illness and this can impact upon the way a young person responds to chronic illness (Woodgate 1998). Similarities to the way the young people with lupus responded to their diagnosis can be drawn with the findings of teenagers' responses to diagnosis of depression (Wisdom & Green 2004). Wisdom and Green (2004) identified three types of response to diagnosis of depression: 'identity infusers' who intrinsically accepted their diagnosis as part of them; 'labellers' who were relieved at diagnosis and who perceived diagnosis as fundamental to recovery; and 'medicalizers' who adopted the 'patient' role while also concerned /worried at receiving their diagnosis (Wisdon & Green 2004). Drawing on Wisdon and Green's (2004) work some young people in this study were pragmatic and rapidly internalised lupus as part of them. These could be described as 'identity infusers'. Some were relieved and responded positively and these could be categorised as 'labellers'. Others were focused on the medical management /treatment but also appeared fearful about consequences of serious illness and could be described as 'medicalizers'.

I would argue that the responses to diagnosis in young people with lupus appears to be grounded in their preceding experiences of 'emerging illness' and 'seeking help' and it would seem to be important to recognise, acknowledge and address these experiential differences at diagnosis. This is supported by the findings from other studies, for example, illness representations based on the self-regulatory model (Leventhal et al 1992) relate to the way in which information about a health threat such as lupus is received and thus interpreted and can provide clues toward how a person is coping with their diagnosis of lupus and indicate the need for enhanced support (Goodman et al 2005). The role of a paediatric psychologist has been acknowledged as of particular importance in enabling young people to adapt and cope with this lifelong chronic disease (Morgan 2013).

Diagnosis was for all young people a significant event and this finding has resonance with findings from other studies such as Charmaz (1991) and it was remembered by the majority of young people as a single moment in time, as also seen in other studies (Charmaz 1991, Kralik et al.2000). This notion of a single point in time contrasts sharply with the evidence presented by Price and Walker (2014) in their qualitative 'virtual focus group' study with adults. In their findings they argue that in lupus there is not a 'diagnostic moment' but a process or journey which for many of their study participants did not result in a diagnostically satisfying conclusion. These contrasting experiences are worthy of note and might be explained due to a number of factors. Firstly, as previously stated, young people differ biologically, emotionally and socially from their adult counterparts. Secondly lupus in young people is characterised by more active disease at presentation than adults (Brunner et al 2007) and thirdly in Price and Walker's (2014) blog-based study it was not known if the participants had a confirmed diagnosis of lupus. Furthermore, just as the experiences of adults contrasted with those of young people, the journey at diagnosis was unique to each young person. Concurrent 'impact on self' was created through negotiations between illness experience and illness representations.

The impact of diagnosis affected both young people and family and this is consistent with literature on chronic illness (Travis 1976, Suris et al 2004, Yeo & Sawyer 2005). The main sources of stress on families were fragmentation of the family unit (the young person with lupus experienced hospitalisation away from home and were accompanied by a parent) and the subsequent impact of this fragmentation had upon families resulting in a burden of care placed particularly on elder siblings. In addition, there was evidence of parental guilt due to separation from their home and family; this was the result of an increased requirement for the young person and accompanying parent to remain in hospital for substantial periods of time because of a severe and difficult to manage form of lupus. This separation from home further compounded feelings of parental guilt as younger/other siblings suffered as a consequence and they had to manage without their main carer/parent. Hospitalisation was intrinsically linked to diagnosis in the young peoples' experiences as a 'variable' that did not exist prior to diagnosis, was intrusive to family functioning and required their negotiation. Young people showed maturity and insight into the far-reaching effects of their diagnosis. They acknowledged their journey through reflections in which they placed their family as central to their diagnosis, illness experiences and illness representations.

### Resilience, Reflection & Recovery

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All of the young people stories of their 'journey to diagnosis' were constructed through their experiences which were contextualised by their 'lupus present' and 'lupus future'. It would be impossible to remove the influence of hindsight from their reflective accounts and I would argue that this would be detrimental to the findings. Fundamentally, young people needed to experience life beyond diagnosis to colour in the detail, it was this viewpoint that enhanced perception, added focus and ultimately differentiated between illness and recovery.

The young people revealed evidence of 'resilience' in their acceptance, adaptability and coping with diagnosis. This is a concept much documented in the literature (Ahern 2006, Alvord & Grados 2005, Brooks 2006, Masten 2011, Olsson et al 2003, Zolkoski & Bullock 2012) and is a characteristic recognised in young people when faced with adversity as they are often able to successfully overcome challenge. The psychosocial strength of young people is apparent when challenged by juvenile rheumatoid arthritis in Noll et al.'s (2000) study where maximum treatments impose huge restrictions upon life and functioning. For young people with lupus much of their resilience and their ability to cope is contextualised and illustrated by whether they are able to attend school and their ability to participate, socialise and achieve academic success. Reduced social functioning and isolation is evident in the young people with lupus, this concurs with both young people with chronic illness (Meijer et al 2000), and adult populations with lupus (Woodgate 1998).

It is evident from the findings of my study that diagnosis impacted on young people and their families in different ways. Some young people responded pragmatically, able to accept lupus quickly and normalising lupus in their lives, while others did not. Marshall et al. (2009) identified the concept of children with diabetes and their parents placing a disease in the background in an effort to live a 'normal life'. As with parents in Marshall's theory of 'normalisation', the parents of the young people with lupus used similar strategies to ensure continued family functioning and strengthening of the family unit. In young people with lupus this normalisation was also apparent in their reflections looking back upon diagnosis. They were able to contextualise when they felt sick before diagnosis and compare it to feeling well and 'normal' during the recovery stage. Medication was integral to recovery for young people with lupus and they perceived a successful recovery as attainable through adherence to their medication regime.

The young people reflected upon their 'journey to diagnosis' as being lengthy and this is unsurprising as similar findings have been identified in relation to adults' experiences of diagnosis

(Waldron 2012, Price & Walker 2014). A lengthy time to diagnosis was identified by the majority of young people as having a negative impact on their experiences of lupus. They would rather have experienced a speedier time to diagnosis and they perceived a reduction in the time taken from onset of symptoms to diagnosis would have been beneficial.

## Passing of Time

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The concept of time in chronic illness has been well established (Charmaz 1991, Fisher, 1982, Ostroska 2008) and the young people with lupus experienced its relevance as it defined, demarcated and added perspective to their 'journey to diagnosis'. Furthermore time was experienced differently during the different stages of their illness as time was not a static concept. During 'emerging illness' time passed slowly, and elasticity was observed as the boundaries of time during onset of symptoms appeared indistinguishable and fluid. Whilst 'seeking help' time continued to pass slowly but was abbreviated by symptoms of illness which took precedent over daily life. Passing of time was defined by episodes of seeking help that failed to negate health concerns. At diagnosis, time almost stood still for a single moment for the young people as they reached the pinnacle of their journey whence a definite diagnosis was made. Some young people experienced a rapid quickening of time as diagnosis overwhelmed and engulfed their lives. Others adopted a more pragmatic approach and time started to normalise as they adapted to their new diagnosis with relief and assertion. Time after diagnosis is on-going for the young people. For some young people it is strongly influenced by their lupus experience and time is expressed 'since diagnosis' whilst others have sought to place lupus in the background and time exists in the present sense.

Fundamental to the young people's experiences of time is that the 'passing of time' adds meaning and boundaries to their experience of their illness, diagnosis and treatment. Time is also reflected as a continuum whereupon timeframes can merge together to form the complete experience (Charmaz 1991); this prevented young people remaining static in one experience. Illness

represented by a trajectory displaying several segments or timeframes has been postulated (Glaser & Strauss 1968) and further developed by Reimann and Schutze (1991) who suggested that experiences are not subject to a singular but many overlapping trajectories. The illness trajectory had been described as having three stages: firstly the time needed to make a diagnosis; secondly adjustment to the illness and thirdly the terminal phase. It has been argued that within these three segments different trajectories co-exist representing different patient experiences (Ostrowska 2008). The findings of this study resonate with Reimann and Schutze's (1991) staged approach with the period of emerging illness, and seeking help, preceding stage one. Stage one broadly aligns with young people's experiences of diagnosis and elements of stage two are present in resilience, reflection and recovery.

## Limitations

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As a researcher-practitioner I have been aware of how I may have influenced and potentially limited the data. As a children's nurse and research nurse working with children and young people with lupus, I had knowledge of their condition and I would have held some preconceived ideas about the disease trajectory and what to expect. At the beginning of this study I was new in post having previously worked with children and young people with arthritis. I was keen to learn from the young people about their experiences with lupus but fully acknowledge the significance of my professional role.

The way I approached and designed the study and the questions I asked will have shaped the data. For example, by asking the young people to tell me their story from 'symptom onset to diagnosis', I artificially imposed a beginning and end point to their story. I reflected on whether I made the time point of diagnosis significant by the nature of my questioning. I would argue that, as

the findings reveal, the young people were eager to tell their stories beyond diagnosis and were able to contextualise their experiences by reflecting on their past and future with lupus.

Other limitations include aspects such as the scale of the study; it was a small exploratory study involving only eight young people who had received their diagnosis of lupus after/on 1st January 2010. Although qualitative research does not aim to generalise, the small scale does limit the conclusions that can be drawn and the generalisability of the findings. Also, there was only one male participant which means the study reflects the experiences of young women rather than young men; however, the fact most participants were female is generally reflective of the female preponderance of the disease.

I was also part of the multidisciplinary team as a research nurse caring for all the young people and was present at half of the consultations with young people when a diagnosis was given. This gave me insight and information that I otherwise would not have had. I had to work hard when analysing my data to ensure that I only worked with the data I had collected rather than impressions formed during my clinical time with them. Although this potentially might have affected reliability and rigor I felt it actually enhanced data collection and concurrent analysis. The young people had already established a relationship with me and I felt this facilitated the interview conversation rather than hindered it. I was able to maintain my position as a researcher rather than research nurse by conducting the interview in my own clothes, away from the clinic area and emphasising my role in the study. Being present at four of the diagnosis giving experiences was pivotal to my analysis as the stories the young people told contrasted with my own recollections, thus highlighting the differences for young people and professionals in such consultations. It heightened attention to detail, acting as motivation and inspiration to listen to the stories the young people tell about their experiences of diagnosis.

## Recommendations for Practice

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The recommendations for practice based on the findings of this study are that:

1. Since young people may not recognise symptom-related changes, support should be directed towards facilitating and educating young people to recognise such changes.
2. Mechanisms, especially within primary care for young people and families seeking help for such symptoms, should be improved by education of both primary care providers and young people and families about the signs and symptoms of JSLE.
3. Awareness should be raised within the wider community, including schools, of emerging symptoms of lupus in young people by health education campaigns.
4. All young people should have the opportunity to reflect and tell their story from 'symptom onset to diagnosis' to an appropriate member of the healthcare team. The perceived advantages of this are allowing young people to detail their experience from their perspective, enabling the disclosure of things that were important to them and things that went well and things that did not. In the future as we move towards personalised healthcare and facilitating the transition of a young person to adult services this may be of particular relevance. This process might be best achieved without a parent or family member present to ensure the young person's perspective is fully acknowledged.
5. Increased support, especially at school, should be available for young people after they have been diagnosed.
6. On-going support provided by the multi-disciplinary team (including psychology and the school nurse) and families beyond diagnosis is crucial for young people to enable them to successfully negotiate this life long illness through adolescent transition into adulthood.

## Recommendations for Research

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The recommendations for research based on the findings of this study are that further qualitative studies are indicated into the experiences of young people with lupus:

1. A longitudinal prospective study following young people from point of diagnosis through to transition into adult services utilising a qualitative mixed methods approach using in-depth interviewing and a nominal group approach. This would span critical time-points for a young person with lupus.
2. A multicentre study to compare experiences of diagnosis in young people across the UK to determine the similarities and differences in responses/approaches.
3. A multicentre research study to explore the symptoms of lupus experienced by young people with the aim of developing tools/strategies to raise awareness of the symptoms of lupus in young people.
4. A longitudinal prospective study comparing the experiences of young people with lupus and the experiences of their primary care practitioners (including school nurses) involved in their care: from diagnosis over a one year period.

## Conclusion

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The challenge to specialist rheumatology teams in providing holistic care to young people with lupus is considerable. Crucially the young people in this qualitative study have enhanced our knowledge about the period leading up to, and including, diagnosis from their unique perspective providing insight into issues that are important to them. This is of particular relevance in lupus when improved access to care and earlier diagnosis could significantly help reduce the impact and burden of this, at present, incurable disease. Further qualitative studies are indicated into the experiences of young people with lupus to explore issues important to them, improve outcomes and facilitate standards of care. Understanding young people's experiences of 'symptom onset to diagnosis' has the potential to reduce the impact and burden of this disease.

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## APPENDICES

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## Appendix 4



## Appendix 5

