# ‘Make Every Child Count’
# Estimating current and future prevalence of children and young people with life-limiting conditions in the United Kingdom

**FINAL REPORT**
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# List of abbreviations

| | |
|---|---|
| CI | Confidence Interval |
| CIPS | Continuous Inpatient Spells |
| CNS | Central Nervous system |
| GOR | Government Office Region |
| HES | Hospital Episode Statistics |
| ICD-10 | International Classification of Diseases version 10 |
| LLC | Life-limiting Condition |
| LTC | Life-threatening Condition |
| LOS | Length of Stay |
| LSOA | Lower Super Output Area |
| NHS | National Health Service |
| ONS | Office for National Statistics |
| RESGOR | Government Office Region of Residence |
| SD | Standard Deviation |

# Foreword

Good data is a prerequisite for the planning and evaluation of any health intervention. For many years we have had excellent disease-based registries for cancer and diabetes, and these have been vital in improving services. Regrettably, not so for other potentially life-limiting diseases, of which there are many, and where the situation is often far more complex. There is no clear definition which captures all conditions; hence a wider approach has to be taken to data collection, using disease codes which are part of routine national data collection for health service episodes.

Lorna Fraser and her team have done an excellent job using such routinely collected health data and codes to estimate both the current and future prevalence of life-limiting conditions in children and young people in the UK. One of the major strengths of this study is that they have used the same methodology as in their previous study eight years ago and this brings real added value to the interpretation of the current findings.

There is undoubtedly an increasing prevalence, rising from 52,633 to 86,625 over the last 10 years. This is at least partly due to increasing survival. With advances in medicine and improved care many children who would have previously died in infancy are now living into adolescence, and in some cases even adulthood.

Within the group of children with life-limiting conditions, 2000 will die each year. These children and their families will need access to a very wide range of professionals and services across hospital and community, as well as support from other statutory and voluntary agencies. Many will also need access to specialist palliative care and / or hospice services, both of which are under increasing workforce and financial pressures.

However, it is equally important to recognise that a majority of children with LLC are able to lead active and fulfilling lives for many years, provided that they have access to the care they need to keep them well. This means that all those working in paediatric services in all specialties and locations, need to be trained and equipped to provide the best possible care.

The distinction between severe disability, complex health needs and palliative care is becoming increasingly less relevant, and children with life-limiting conditions are everyone's business. This has major implications for service planning, as well as for the training of all those working in children's services. Stresses on families and siblings need to be recognised, and integrated service planning and commissioning across health, education and social care are essential in order to optimise the health and wellbeing of the entire family, not just that of the young person with a life-limiting condition.

We are grateful to the True Colours Trust for funding this research and for their ongoing commitment to a very important and somewhat Cinderella part of our health system.

**Professor Sir Alan Craft**

President of Together for Short Lives

Emeritus Professor of Child Health

**Dr Hilary Cass OBE**

Chair of Trustees of Together for Short Lives

## Executive Summary

1. Life-limiting and life-threatening conditions (LLC) are terms which have been used to describe the population of children who may benefit from input from paediatric palliative care services.

2. Palliative care for children and young people with life-limiting or life-threatening conditions is an active and total approach to care, from the point of diagnosis or recognition throughout the child's life and death.

3. This study used routinely collected hospital and death certificate data from England to provide an update of current numbers and prevalence of children (0-19 years) with a life-limiting condition and estimate future prevalence (up to 2030).

4. Due to data availability, estimates of future prevalence for Scotland, Wales and Northern Ireland were based on data from Scotland.

5. Children were identified as having a LLC using a list of previously developed diagnostic codes (ICD-10). The dataset for England contained 359,643 individuals over a 17 year time period.

6. The number of children with a LLC identified in this dataset from England rose from 32,975 in 2001/02 to 86,625 in 2017/18. Excluding some diagnoses which may not be considered LLC reduced this number to 81,712 in 2017/18.

7. The national prevalence of LLC in children (aged 0-19 years) in England had increased over 17 years from 26.7 per 10000 in 2001/2 to 66.4 per 10000 in 2017/18. Excluding some diagnoses which may not be considered LLC reduced this prevalence only slightly to 63.2 per 10000 in 2017/18.

8. There was some evidence in these data that this increase in prevalence was driven by both an increase in recording of these diagnoses and an increase in survival in this population. The former may reflect a change in coding practice rather than a true increase in incidence.

9. The prevalence of LLCs was highest in the under 1 year age group and increased from 130.1 per 10,000 in 2001/02 (n=7255) to 226.5 per 10,000 in 2017/18 (n=15,489).

10. The prevalence of LLCs was highest for congenital abnormalities which by 2017/18 was 27.2 per 10,000 more than twice the next most prevalent group, neurological disorders (10.8 per 10,000).

11. The prevalence of LLCs was significantly higher among boys (72.5 per 10,000 vs girls 60.0 per 10,000 (2017/18)) although there was no difference in the rise in prevalence between sexes over time.

12. Prevalence of LLCs was highest amongst children of Pakistani origin (103.9 per 10,000) and lowest among children of Chinese origin (32.0 per 10,000) in 2017/18. This is important in terms of flexibility of service to meet the needs of all children.

13. More children than expected with a LLC lived in areas of higher deprivation (13% most deprived versus 8% in least deprived). The deprivation categories were population weighted therefore you would expect ~10% of children to have a LLC in each category.

14. The future prevalence of children aged 0-19 years with a LLC in England is estimated to be between 67.0 and 84.2 per 10,000. There is a range of uncertainty around these estimates.

15. The estimated future prevalence in 2030 for Scotland (51.0-55.8 per 10,000), Wales (50.8-55.6 per 10,000) and Northern Ireland (52.6-56.5 per 10,000) are lower which may reflect different demographics of the population.

16. 10.4% (n=37,328) of these children with a LLC died during the study period, 8.4% (n=30,187) of whom died before age 20. There are a large number of deaths both in those under 1 year of age but also in young adults, highlighting the need for age and developmentally appropriate services.

17. There are increasing numbers of children with a LLC who have a hospital stay of greater than 28 days each year, rising from 2482 in 2001/2 to 3538 in 2017/18. This will impact on hospital services.

18. These data did not contain any measure of complexity of the underlying condition or the needs of the child or family, future research and data collection should address this gap.

# Background

Life-limiting and life-threatening conditions are terms used to describe the population of children who may benefit from input from paediatric palliative care services (1, 2). Life-limiting conditions (LLC) are those for which there is no reasonable hope of cure and from which children or young people are expected to die (3). Life-threatening conditions (LTC) are those for which curative treatment may be feasible but can fail, such as cancer (3). This population of children with life-limiting and life-threatening conditions (hereafter referred to as LLCs), is a very heterogeneous group with nearly four hundred individual diagnoses classified as life-limiting or life-threatening (4).

Paediatric palliative care differs from adult palliative care in that the World Health Organisation recommends it begins when a condition is diagnosed and continues regardless of whether a child is treated for the disease or not (5). This means that children and their families may require care and support for a prolonged period of time; more than 20 years in some instances (6). Palliative care for children and young people with life-limiting or life-threatening conditions is an active and total approach to care, from the point of diagnosis or recognition and throughout the child's life and death. It embraces physical, emotional, social, and spiritual elements, and focuses on enhancing quality of life for the child/young person and supporting their family. It includes the management of distressing symptoms, provision of short breaks and care through death and bereavement (3).

The number of children with a LLC has been rising with an estimated increase of 29% in the decade up to 2009/10 and a reported 40,000 children and young people having a LLC in England (7). More recent estimates from Scotland show that the prevalence of children and young people with a LLC has continued to increase and that if estimates are based solely on children with a hospital admission in a year then these data were an underestimation of the true number (8). Previous studies have also indicated that prevalence varies by ethnicity, deprivation and geographical region (7-9).

Over the last 30 years, there has been an increase in the number of paediatric palliative care and hospice services in the UK that provide palliative and end of life care for children, but there is little evidence on the models of care, quality, resource implications and outcomes of children and families who use these services. We know that these services vary in their professional configuration, services provided, funding sources and population served (3). These services have developed locally with heavy reliance on individual clinician and third sector organisations e.g. children's hospices (10). As a result, delivery of palliative care for children is 'inconsistent and incoherent' (4). Many of these children are also cared for across paediatric specialities including community paediatrics (11). Planning for development of current and future services is difficult without up to date data on the population of children and young people who would benefit from these services (12).

## Aim

To estimate the current national prevalence of children with a LLC in the UK and develop a model to predict the future prevalence of children with a LLC in the UK (2017- 2030).

### Objectives

1. To assess trends in prevalence of children with a LLC (by age group, ethnic group, deprivation, Government Office Region (GOR)(2001/02-2017/18) using hospital and death certificate data for England;
2. To model future national prevalence of children with a LLC in the UK utilising ethnic specific population projections (up to 2030);
3. To model future subnational prevalence of children with a LLC utilising ethnic specific population projections (up to 2030).

# Methods

## Data sources

### England

An extract of Hospital Episode Statistics Admitted Patient Care (HES) linked to the Office for National Statistics mortality data were obtained from NHS Digital (13). These data include information on all admitted care in NHS hospitals in England whether it be planned or emergency, overnight stays or day cases, patients resident outside England or care delivered by treatment centres funded by the English NHS. HES records include information about clinical diagnosis and procedures, patient information including age, sex and ethnicity, dates of admission and discharge and geographical information such as local authority of residence (14).

### Scotland, Wales and Northern Ireland

Data from a previous study were available at the individual level from 1 April 2003 to 31 March 2015 for Scotland. These data were used for current estimates but not reanalysed.

Data were available from 1st April 2001 up to 31st March 2010 for Wales and Northern Ireland, but only at an aggregated level. Current and future estimates for Wales and Northern Ireland were based on trends in Scotland as these were the closest comparable data available at the individual level.

## Life-limiting conditions

LLCs were defined according to a list of previously developed ICD-10 codes (7, 15) which was devised through a number of steps (Figure 1). Firstly, a list was developed using two independent sources of information: the Hain Dictionary (16) version 1.0 of ICD-10 codes for children seen by palliative care providers and a list of diagnoses for children accepted for care at Martin House Children's Hospice, Yorkshire, England from 1987 to 2010. A 4-digit ICD-10 code was assigned to 92% of diagnoses on the Martin House list; the 8% not coded were children without clear diagnoses (e.g., "degenerative neurologic disease with no firm diagnosis"). Combining both sets of codes produced a provisional list of 801 ICD-10 codes for

further scrutiny (84% of the codes appeared on both lists). All of these ICD-10 codes were individually subjected to the following two questions:

1) Are most children with this diagnosis life-limited/life-threatened?
2) Are most sub-diagnoses within the ICD-10 code life-limiting/life-threatening?

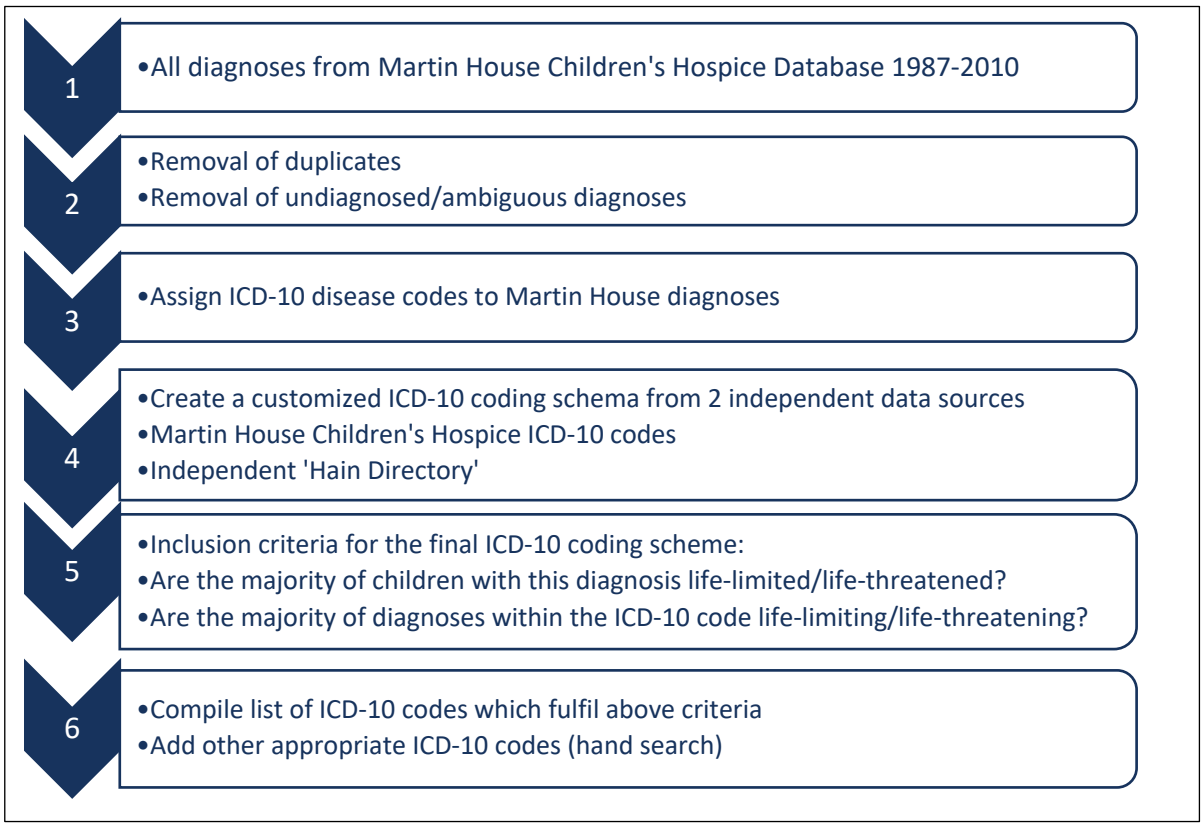


***Figure 1: Flow diagram of the development of the ICD-10 coding framework***

A list of ICD-10 codes that fulfilled these criteria was compiled and completed by adding all malignant oncology ICD-10 codes. The final ICD-10 coding framework consisted of 777 4-digit ICD-10 codes (Table 1). Malignant oncology codes accounted for 445 (57%) codes, with congenital malformations and chromosomal abnormalities having 87 (11%) codes.

This ICD-10 coding framework has been shown to be sensitive (i.e. it identifies the children with a LLC) by identifying 75% of children who died in paediatric intensive care units (17) but there are some concerns about its specificity (i.e. is it also picking up children who do not have a LLC). This is due to the grouping of diagnoses within ICD-10 and the variation in clinical features of some of these diagnoses. Therefore, in this study an attempt was made to refine this list, as described below:

The list of ICD-10 codes was assessed by the independent advisory panel for this study and a group of codes/exclusions were identified where the panel felt that the child may not be

always be considered as having a LLC or LTC. These codes, each fell into one of the following categories;

I. Perinatal diagnoses beyond the age of 1 year. Restricting inclusion of perinatal diagnoses to age under 1 (to be included beyond age 1 a non-perinatal LLC diagnosis is required)
II. Oncology cases 5 years after first oncology diagnosis (assuming no other LLC is present)
III. Non central nervous system (CNS) oncology cases 5 years after first oncology diagnosis (assuming no other LLC is present)
IV. Early stage (1-3) renal failure (only appeared in post 2010 ICD classification)

A sensitivity analysis assessing the impact of removing these codes was undertaken.

**Table 1: ICD-10 diagnostic coding framework used to identify and categorise children with life-limiting conditions.(7)**

| Diagnostic Group | ICD-10 Numbers |
|---|---|
| **Neurology** | A17 A810 A811 F803 F842 G10 G111 G113 G12 G20 G230 G238 G318 G319 G35 G404 G405 G600 G601 G702 G709 G710 G711 G712 G713 G800 G808 G823 G824 G825 G934 G936 G937 |
| **Haematology** | B20 B21 B22 B23 B24 D561 D610 D619 D70 D761 D81 D821 D83 D891 |
| **Oncology** | C D444 D48 (Central Nervous System: C70,C71,C72, D33, D43) |
| **Metabolic** | E310 E348 E702 E71 E72 E74 E75 E76 E77 E791 E830 E880 E881 |
| **Respiratory** | E84 J841 J96 J984 |
| **Circulatory** | I21 I270 I42 I613 I81 |
| **Gastrointestinal** | K550 K559 K72 K74 K765 K868 |
| **Genitourinary** | N17 N18 N19 N258 (Early stage (1-3) renal:N181, N182, N183) |
| **Perinatal** | P101 P112 P210 P285 P290 P293 P350 P351 P358 P371 P524 P525 P529 P832 P912 P916 P960 |
| **Congenital** | Q000 Q01 Q031 Q039 Q040 Q042 Q043 Q044 Q046 Q049 Q070 Q200 Q203 Q204 Q206 Q208 Q213 Q232 Q218 Q220 Q221 Q224 Q225 Q226 Q230 Q234 Q239 Q254 Q256 Q262 Q264 Q268 Q282 Q321 Q336 Q396 Q410 Q419 Q437 Q442 Q445 Q447 Q601 Q606 Q614 Q619 Q642 Q743 Q748 Q750 Q772 Q773 Q774 Q780 Q785 Q792 Q793 Q804 Q81 Q821 Q824 Q858 Q860 Q870 Q871 Q872 Q878 Q91 Q920 Q921 Q924 Q927 Q928 Q932 Q933 Q934 Q935 Q938 Q952 |
| **Other** | H111 H498 H355 M313 M321 M895 T860 T862 Z515 |

## Patient data

An extract of clinical and demographic information on all hospital episodes for children between the ages of 0-19 years who had ever had an ICD-10 code for an LLC (Table 1) recorded within the admitted patient HES was received from NHS Digital. These data were available for the financial years beginning 1st April 2000 until 31st March 2018.

These HES data were linked to the Office for National Statistics (ONS) death certificate data and, if the child had died, information on date of death and cause of death was available (13).

## Population data

Population estimates broken down by age, sex, ethnicity and Government Office Regions were obtained from http://ethpop.org (18). This source has been used in preference to the sub-national estimates produced by the Office of National Statistics because the cohort component population estimate model (19) incorporates more detailed demographic information by ethnic group in relation to newborns, mortality, and most importantly, both subnational migration and international migration. These data were available as mid-year estimates for 2001-2017 and projected estimates up to 2030. Ethpop includes subnational projections of population by ethnic group, age and sex beyond mid-century based on midyear population estimates for 2001 and 2011. Here we incorporate populations projected to 2030.

## Data cleaning

Hospital episodes for children who resided outside England (identified by Government Office Region code) or for those who were older than 19 years of age were removed from the extract. Data on infants who were recorded as still born using the HES definition of 'a still birth is a birth after a gestation period of 24 weeks where the baby shows no sign of life when delivered' (20) were also removed from the data extract. The financial year in which an LLC was first diagnosed for each child was identified. Hospital episodes occurring prior to this point were excluded for the prevalence analyses but demographic data was used.

**Age -** age was taken from the start age of the first hospital episode in each financial year and grouped into age categories (< 1 year, 1 to 5 years, 6 to 10 years, 11 to 15 years, 16 to 19 years).

**Diagnostic group -** Diagnoses were grouped according to eleven diagnostic groups (neurology, haematology, oncology, metabolic, respiratory, circulatory, gastrointestinal, genitourinary, perinatal, congenital and other) which were mostly based on ICD-10 chapters (Table 1) (7). For the prevalence analyses individuals were allowed to have more than one LLC diagnostic group.

For analyses of the death certificate data a main diagnostic group was determined. Where a child had multiple LLCs that fell into multiple categories, the most frequently recorded category (by hospital episode) was used. In the event of a tie, progressively earlier admissions

were ignored until the tie was resolved.

**Sex -** was recorded as male or female. Individuals with conflicting multiple coding were assigned the most commonly reported sex.

**Ethnicity -** Self-reported ethnicity for each hospital episode was coded according to the 2001 Census groups (21). Eight ethnic groups were made by collapsing the 16 Census groups into the following categories White (White: British, White: Irish, Other White), Black (Black or Black British: Black Caribbean, Black or Black British: Black African, Black or Black British: Other Black), Indian (Asian or Asian British: Indian), Pakistani (Asian or Asian British: Pakistani), Bangladeshi (Asian or Asian British: Bangladeshi), Chinese, Mixed (Mixed: White and Black Caribbean, Mixed: White and Black African, Mixed: White and Asian, Mixed: Other Mixed), other Asian. Conflicting, non-missing, multiple ethnicity was assigned the most commonly recorded ethnicity, ties were left unresolved (n=3376 (0.8%)).

**Government Office Region -** The nine Government Office Regions (GOR) of residence were used as subnational geographical areas North East, North West, Yorkshire and Humberside, East Midlands, West Midlands, East of England, London, South East, South West. In cases of multiple GORs the first GOR per financial year was used. GORs coded as "Unknown" were replaced with the first known GOR for that year.

**Deprivation -** An index of multiple deprivation (IMD2010) (22) was assigned to each individual based on the 2001 Lower-layer Super Output Area (LSOA) of residence. If there was no known LSOA in a year, but the individual was known from Government Office Region of Residence (RESGOR) to be in England then (in preference order) the last known LSOA from preceding years or the next known LSOA from later years was assigned. Ten deprivation categories were created, from least (category 1) to most deprived (category 10), based on IMD2010 scores. These were population weighted so that each category contained approximately 10% of individuals in England aged 0-19 years. Assignment of deprivation code was undertaken each year and if an individual moved during that year the deprivation code associated with the first LSOA in that year was used.

## Analysis

The number of children with a LLC were identified and counted each year (see case identification below). As population data was only available from 1st April 2001, numbers were only presented from this time point. The prevalence and 95% confidence intervals were calculated per 10,000 population at risk (aged 0-19 years).

$$prevalence = \frac{\text{number of individuals with an LLC}}{\text{population at risk}} \; x\, 10000$$

Prevalence was calculated as an overall total and stratified separately by age group (including a comparison between infants (<1 year of age) and non-infants), disease category (also including a comparison between infants and non-infants), sex, ethnic group, government office region and level of deprivation.

## Case identification

An individual child was included in the prevalence calculations if they fulfilled the following criteria:

1. had a diagnosis of one of the LLC/LTC ICD -10 codes in this year or a previous year (from April 2001);
2. had a hospital admission in the year of analysis[1];
3. were <19 years old;
4. were resident in England.

## Denominator data

The population at risk was estimated using ethnic specific population data sourced from the ETHPOP dataset (18).

95% confidence intervals (CI) for the prevalence estimates were calculated using standard methods for CIS for proportions (23).

## Sensitivity analysis

A series of sensitivity analyses were conducted where the definition of an LLC was restricted to exclude the following four sets of diagnoses identified by the advisory board, individually and combined to assess the effect on overall prevalence figures:

(i) Perinatal disorders were assumed not to be relevant after the first birthday[2];

(ii) Oncology cases 5 years after diagnosis after which point they were assumed to be resolved;

(iii) Non-central nervous system (CNS) oncology cases 5 years after diagnosis after which point they were assumed to be resolved;

(iv) Early stage (1-3) kidney failure: hospital admissions for these cases were not included when no other LLCs were present.

[1] Previous research from England only included children in a year if they had a hospital admission for one of the LLC codes

[2] Assumption is that if they had an ongoing LLC after age 1 this would be recoded e.g. a baby with severe birth asphyxia would be recoded as having cerebral palsy

### Assessment of change in prevalence

Additional analyses were undertaken to explore whether the change in prevalence was being driven by a change in incidence or a change in survival of this population.

#### Change in incidence:

The following analyses were carried out to analyse any evidence of change in incidence of these diagnoses.

a. The number of new diagnoses (i.e. first recording of an LLC for a particular child) per year, overall and in the under 1s;
b. Trends in the age of first record of diagnoses in these data per year;
c. Change in the number of hospital admissions and therefore increased opportunity for children to be recorded within these diagnoses.

#### Change in survival:

The following analyses were carried out to examine changes in survival.

a. The number of children who died overall and per financial year, stratified by age and main diagnostic group;
b. Proportion of the children who died each year;
c. Median age at death per year;
d. The probability of survival per year since the 1st LLC diagnosis (using a smaller birth cohort, n=237,210).

Due to these data being left censored, a traditional survival type analyses could not be undertaken as there was no information on children who died prior to 1998 and therefore it was not possible to construct a birth cohort retrospectively (24). However, it was possible to construct a birth cohort of all children born since 2001 and estimate their probability of survival until 2017 using Kaplan-Meier analysis.

In some instances, death may be referred to the Coroner which can result in delays of up to three years for a death to be recorded. Therefore, the latter part of the dataset may be underestimating the deaths in this population (24).

## Additional Analysis

### Proxy measure of complexity

In the absence of data on needs and complexity, an assessment was undertaken using long stay in hospital as a proxy measure of complexity. A length of stay of >28 days was defined as a long admission (24). Length of stay (LOS) was calculated by counting the length of continuous inpatient spells (CIPs) per financial year (discharge date - admission date). CIPs represent continuous hospital stays including transfers to other hospitals and are calculated by combining individual finished consultant episodes within HES data. LOS that spanned financial years were split into parts (pre April 1st and post April 1st). The maximum LOS per

financial year was determined for each individual. Individuals were categorised as being a day case (no overnight stay), having an admission of 1-13 nights, 14-27 nights, 28-55 nights or 56 nights or more. Data on whether the child was admitted as an emergency were also available.

In order to analyse factors associated with a long admission (stay >28 days (yes/no)), multilevel (hierarchical) logistic modelling was used to account for the repeated (annual) measurement of maximum LOS, with individuals at level 2 and year at level 1. Year, sex, age group, ethnicity, GOR, deprivation (collapsed into five categories with the least deprived (category 1)), main diagnostic group and emergency admission (yes/no) were all entered as covariates. As there may be additional reasons for a birth admission to be >28 days (e.g. prematurity) a sensitivity analyses excluding birth admission was also undertaken.

### The number of young people who would require adult services each year

The Advisory Committee recognised the lack of provision for young adults and it was decided to calculate the number of young people who were alive in the year they were 19 years old. We also calculated the number of individuals aged over 19 years who died within the study period.

## Modelling of future prevalence

Due to the difference in prevalence amongst the age groups and ethnic groups, future prevalence was estimated using a population based modelling approach (Figure 2) (25). This modelling approach automatically adjusts for changing population demographics and does not require separation of incidence, survival and migration. It requires good population estimates for which Ethpop data were used again (18).

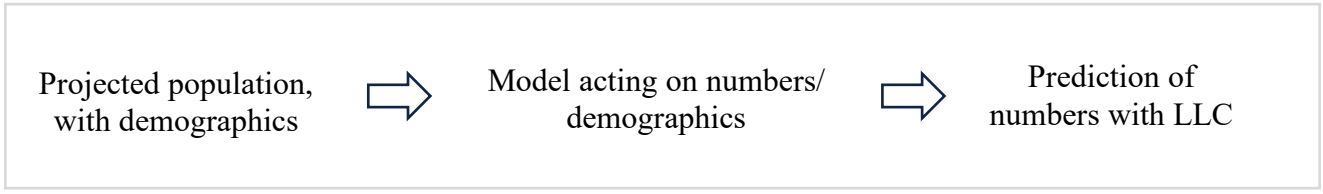


***Figure 2: Future projection modelling approach***

The methods for calculating these future prevalence are detailed in Appendix 1-Methods.

Three models were developed. **Model 1** used estimates of the numbers of individuals with a LLC from 2004-2016 and **Model 2** used estimates from the same time period but where the restricted definition of a LLC was applied (i.e. excluding oncology cases 5 years after 1st diagnosis and perinatal diagnosis 1 year after birth). These two models assume that the rate of change of incidence and survival for children with a LLC would continue however given the uncertainty of any further improvement in survival or increase in incidence, a further model **(Model 3)** was created showing the predicted numbers if there was no further change in survival and incidence.

All data manipulation was undertaken using Microsoft SQL server and statistical analysis using STATA version 15 (Stata Corp, Collage Station, TX). Regression analysis was also performed in STATA with the aid of runmlwin (26) for the multilevel models.

# Results

A total of more than eight million hospital episodes (8,002,959) were included in the initial dataset for 537,940 individuals.

## Data cleaning

Hospital episodes outside of the study period (n=12,094) were removed from the dataset along with 2,303,592 episodes for individuals older than 19 years at the time of the episode (Figure 3). Furthermore, 165,400 episodes were removed as they were for individuals not resident in England. Finally, 981 individuals were removed as they were classified as stillborn (Figure 3).

## Missing data

For most variables there was little missing data. Missing sex (n=2,064 (0.2%)) and deprivation scores (n=3,330 (0.3%)) were excluded from the prevalence calculations split by those characteristics. A total of 29,740 (2.8%) individuals had missing ethnicity. Missing ethnicity was more frequent for the earlier years (7% in 2001/02) and dropped to 2% by 2007/08. Those with missing ethnicity were categorised as a separate group.

## Number of children

The final dataset for analyses contained information on 4,543,386 hospital episodes for **359,643 individuals**.

Table 2 shows the crude numbers overall and by age group. The absolute number of children with a LLC each year rose from 32,975 in 2001/02 to 86,625 in 2017/18.

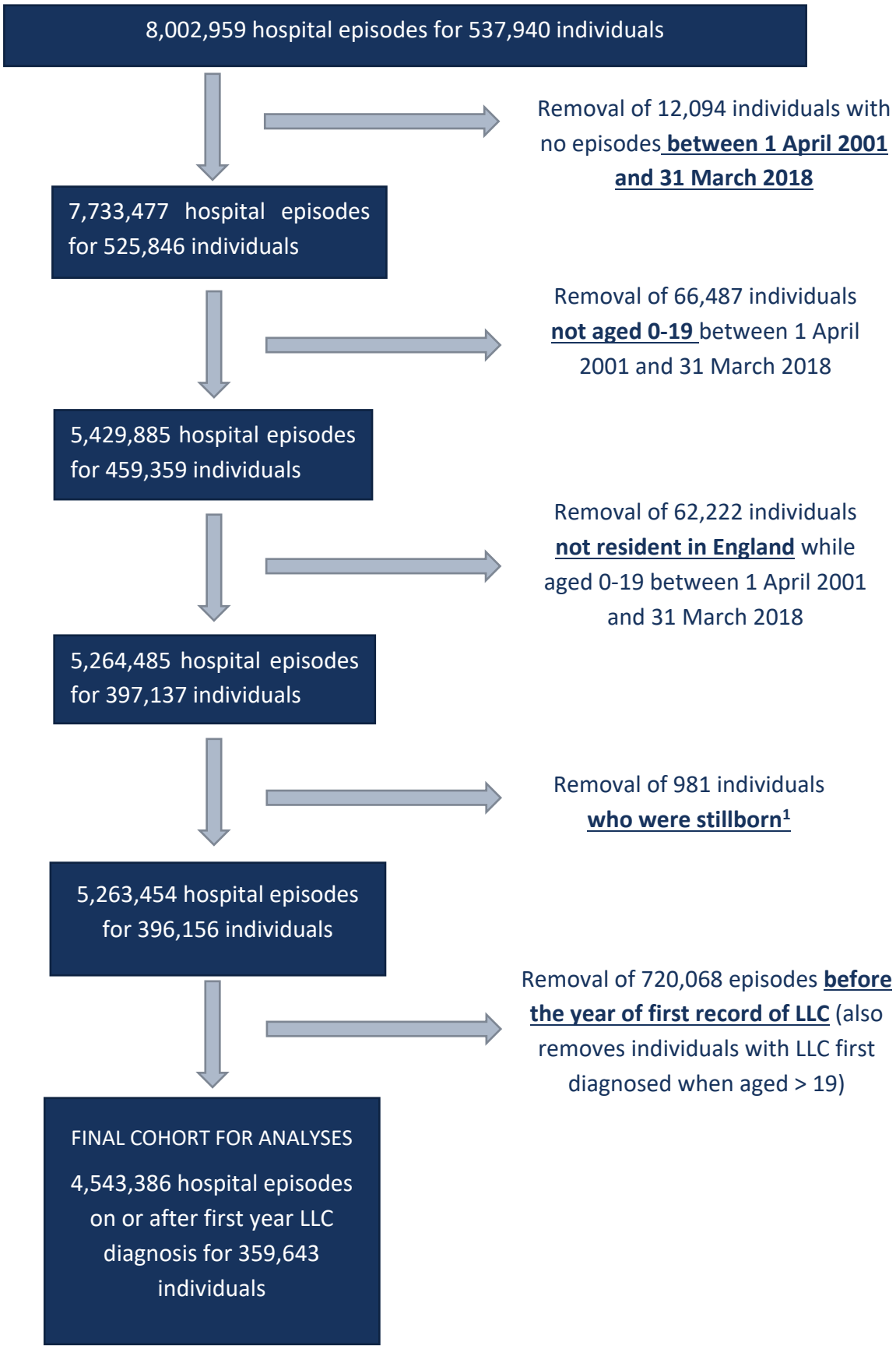


***Figure 3: Flow diagram of inclusion criteria***

[1] Removal of 1031 episodes due to duplicate records

Table 2: Overall numbers and annual prevalence (per 10,000 population) of children (0-19 years) with life-limiting conditions in England by age group for financial years 2001/02 – 2017/18

| Year | Number of patients | Age 0-19 years | Prevalence per 10,000 population | | | | | | | | | | | |
|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|
| | | | 95% CI | Age <1 year | 95% CI | Age 1-5 years | 95% CI | Age 6-10 years | 95% CI | Age 11-15 years | 95% CI | Age 16-19 years | 95% CI | |
| 2001/02 | 32,975 | 26.7 | 26.5-27.0 | 130.1 | 127.2-133.1 | 31.3 | 30.7-31.9 | 19.4 | 18.9-19.9 | 18.7 | 18.2-19.2 | 17.6 | 17.1-18.2 | |
| 2002/03 | 36,688 | 29.7 | 29.4-30.0 | 131.2 | 128.2-134.2 | 37.3 | 36.6-38.0 | 21.8 | 21.3-22.3 | 20.9 | 20.4-21.4 | 19.4 | 18.8-19.9 | |
| 2003/04 | 39,819 | 32.2 | 31.9-32.5 | 134.2 | 131.3-137.2 | 41.7 | 40.9-42.4 | 23.8 | 23.2-24.3 | 22.1 | 21.6-22.6 | 21.5 | 20.9-22.0 | |
| 2004/05 | 42,114 | 34.0 | 33.7-34.3 | 134.4 | 131.4-137.3 | 44.9 | 44.1-45.6 | 25.0 | 24.4-25.5 | 23.3 | 22.8-23.8 | 22.9 | 22.3-23.4 | |
| 2005/06 | 45,974 | 37.1 | 36.7-37.4 | 141.9 | 138.9-144.9 | 48.4 | 47.6-49.2 | 27.8 | 27.3-28.4 | 25.4 | 24.9-26.0 | 25.5 | 24.9-26.1 | |
| 2006/07 | 49,285 | 39.7 | 39.3-40.0 | 158.9 | 155.8-162.0 | 50.5 | 49.7-51.3 | 30.2 | 29.6-30.8 | 26.3 | 25.7-26.9 | 26.8 | 26.1-27.4 | |
| 2007/08 | 52,633 | 42.2 | 41.8-42.5 | 158.7 | 155.7-161.8 | 52.9 | 52.0-53.7 | 33.2 | 32.5-33.8 | 28.5 | 27.9-29.1 | 28.7 | 28.0-29.3 | |
| 2008/09 | 56,436 | 45.0 | 44.6-45.4 | 177.5 | 174.3-180.6 | 54.3 | 53.5-55.1 | 35.5 | 34.8-36.2 | 29.7 | 29.1-30.3 | 30.0 | 29.4-30.7 | |
| 2009/10 | 59,851 | 47.5 | 47.147.9 | 187.2 | 183.9-190.4 | 56.2 | 55.4-57.0 | 38.4 | 37.7-39.1 | 31.5 | 30.9-32.1 | 31.7 | 31.0-32.4 | |
| 2010/11 | 63,256 | 49.9 | 49.5-50.3 | 189.7 | 186.4-193.0 | 58.9 | 58.1-59.8 | 40.7 | 40.0-41.1 | 33.6 | 33.0-34.3 | 33.4 | 32.8-34.1 | |
| 2011/12 | 64,420 | 50.7 | 50.3-51.1 | 174.7 | 171.5-177.8 | 59.9 | 59.1-60.8 | 42.6 | 41.8-43.3 | 35.5 | 34.8-36.2 | 34.6 | 33.8-35.3 | |
| 2012/13 | 69,036 | 54.1 | 53.7-54.5 | 188.1 | 184.9-191.3 | 63.2 | 62.3-64.0 | 45.4 | 44.746.2 | 38.5 | 37.8-39.2 | 36.0 | 35.3-36.7 | |
| 2013/14 | 73,608 | 57.5 | 57.1-57.9 | 212.3 | 208.9-215.7 | 65.7 | 64.8-66.6 | 46.6 | 45.8-47.4 | 41.4 | 40.6-42.1 | 38.5 | 37.8-39.3 | |
| 2014/15 | 77,163 | 60.1 | 59.7-60.5 | 221.4 | 217.9-225.0 | 67.9 | 67.1-68.8 | 48.6 | 47.8-49.4 | 44.5 | 43.7-45.2 | 40.8 | 40.0-41.6 | |
| 2015/16 | 81,172 | 62.9 | 62.4-63.3 | 231.7 | 228.1-235.3 | 71.9 | 71.0-72.8 | 49.6 | 48.8-50.4 | 46.8 | 46.0-47.5 | 42.9 | 42.1-43.7 | |
| 2016/17 | 84,270 | 64.9 | 64.5-65.4 | 235.7 | 232.1-239.3 | 73.1 | 72.1-74.0 | 51.0 | 50.3-51.8 | 49.2 | 48.4-49.9 | 45.9 | 45.1-46.7 | |
| 2017/18 | 86,625 | 66.4 | 66.0-66.8 | 226.5 | 223.0-230.1 | 75.1 | 74.2-76.0 | 52.6 | 51.9-53.4 | 50.9 | 50.1-51.7 | 48.6 | 47.8-49.5 | |

Abbreviations: 95% CI- 95% Confidence intervals

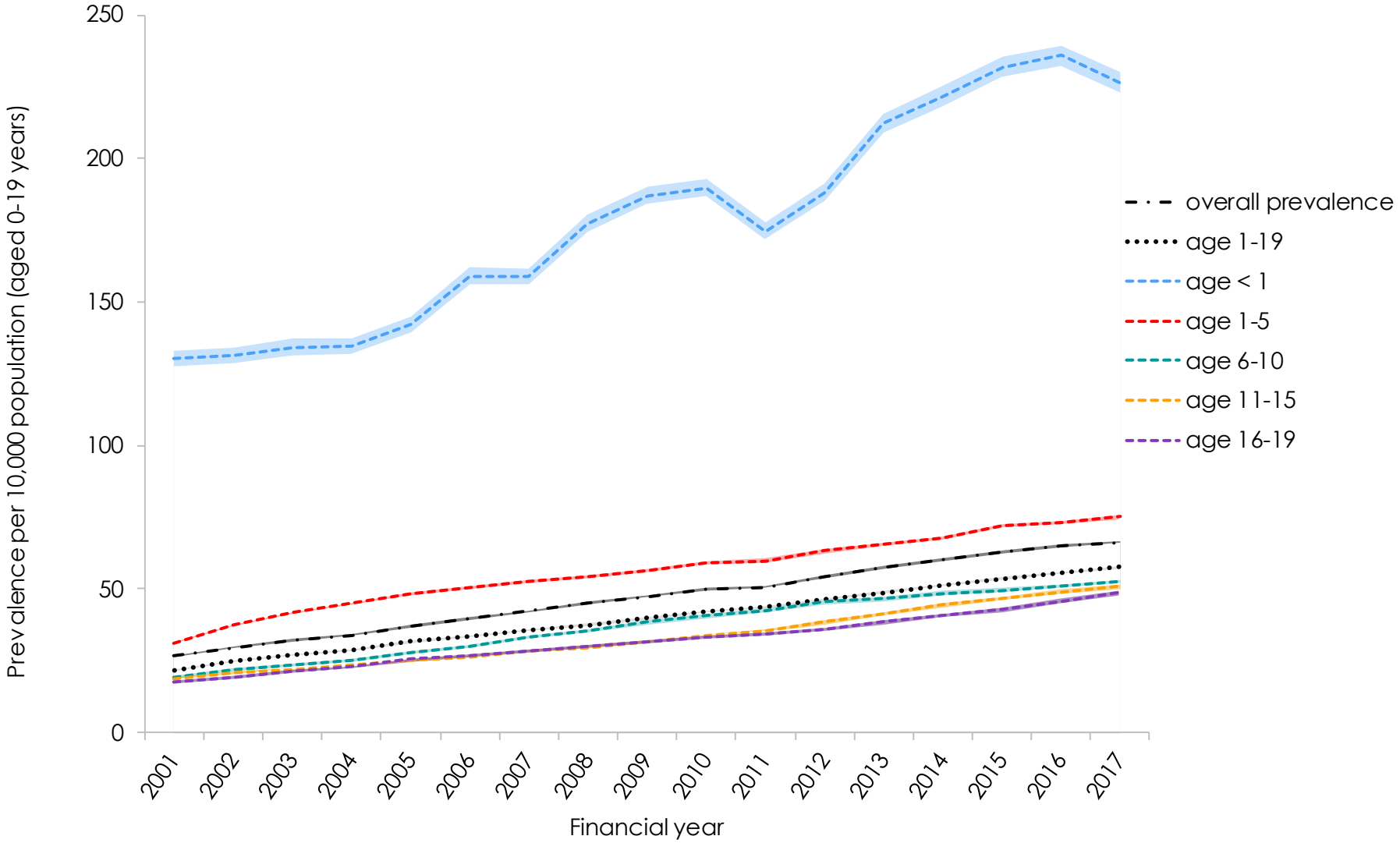


***Figure 4 Prevalence of life-limiting conditions (with 95% confidence intervals in lighter shading) in children (age 0-19) overall and by age for 2001/02-2017/18***

## Prevalence

Table 2 also shows the prevalence per 10,000 population overall and by age group. Overall the prevalence of LLCs has increased from 26.7 per 10,000 (95% confidence interval [95%CI] 26.5-27.0) in 2001/02 to 66.4 per 10,000 [95% CI: 66.0-66.8] in 2017/18 (Figure 4). The prevalence of LLCs was highest in the under 1 age group at 226.5 per 10,000 [95%CI: 223.0-230.1] in 2017/18. The prevalence of LLCs decreased with increasing age but all age groups showed an increase in prevalence over the time period of the study (Table 2).

The increase in prevalence was also largest for under 1-year olds whilst the increase in prevalence was similar in the other age groups (Figure 4). However, the absolute number of children in the under 1 age group is much smaller (average of n~11,500 p.a.) and therefore only contributed slightly to the overall increase in prevalence (Figure 5). When excluding the under 1s, the prevalence for all of the other age groups combined was 21.9 per 10,000 in 2001/02 [95%CI 21.6-22.1] rising to 57.5 per 10,000 in 2017/18 [95%CI 57.1-58.0] (Figure 4).

The number of children with a LLC in more than one diagnostic group increased each year ranging from 15.6% in 2001/02 to 28.4% in 2017/18 (Supplementary Table 1).

The prevalence of LLCs was highest for congenital abnormalities which by 2017/18 was 27.2 per 10,000 (95%CI: 26.9-27.5), more than twice the next most prevalent group, neurological disorders (10.8 per 10,000 [95%CI 10.7-11.0]) (Figure 6). Prevalence was lowest in the circulatory group (2.4 per 10,000 [95%CI: 2.3-2.5]) and the 'other' group (1.7 per 10,000 [95%CI 1.6-1.7]).

There was an increase in prevalence in all diagnostic groups in the study time period, the largest of which was for perinatal disorders, which had a 6-fold increase, and gastrointestinal disorders, which had a 5-fold increase. The smallest increase was among children with a cancer diagnoses (1.6-fold increase). The diagnostic groups differed markedly by age; congenital, neurological and respiratory disorders had the largest increase in prevalence among the 1-19 year olds, whilst perinatal disorders and congenital disorders increased in the under 1s (Supplementary figure 1).

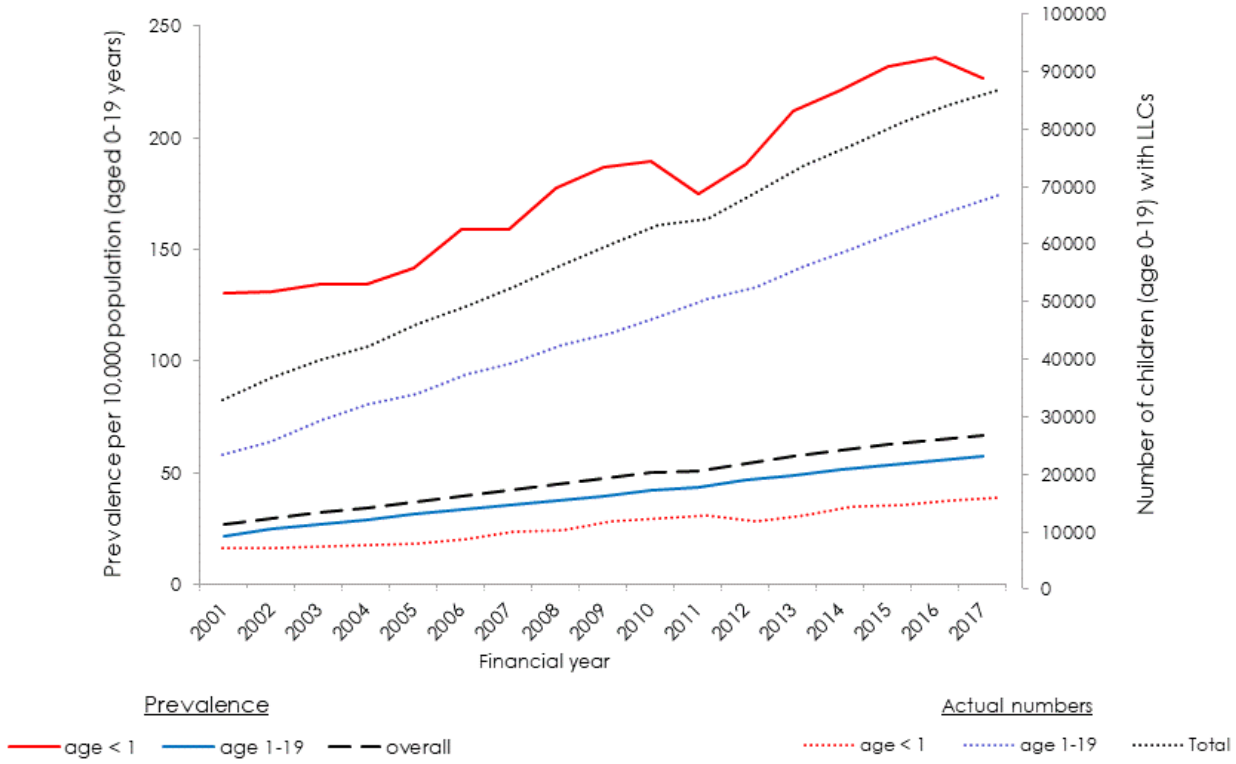


***Figure 5: Comparison of prevalence (and absolute number) of life-limiting conditions between <1 and 1-19 age groups***

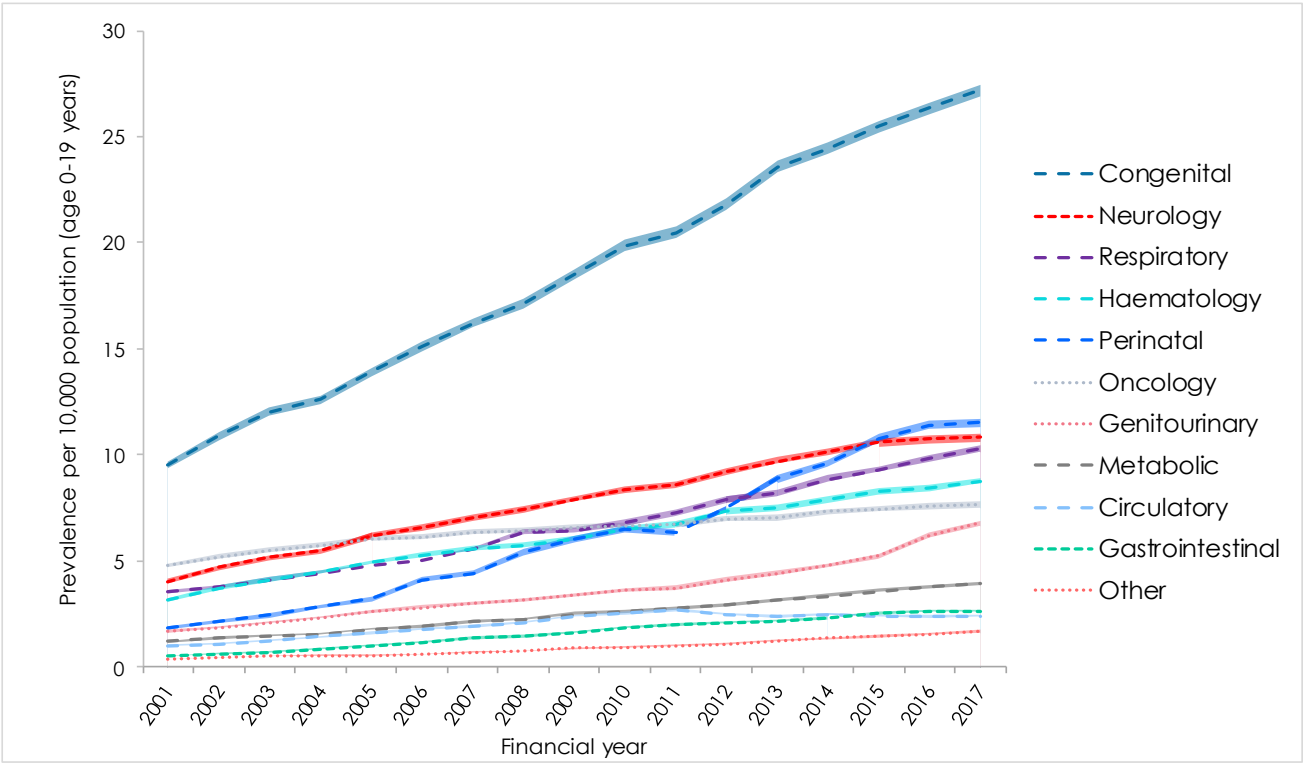


***Figure 6: Prevalence of life-limiting conditions (with 95% confidence intervals in lighter shading) in children (age 0-19) by diagnostic group for 2001/02-2017/18***

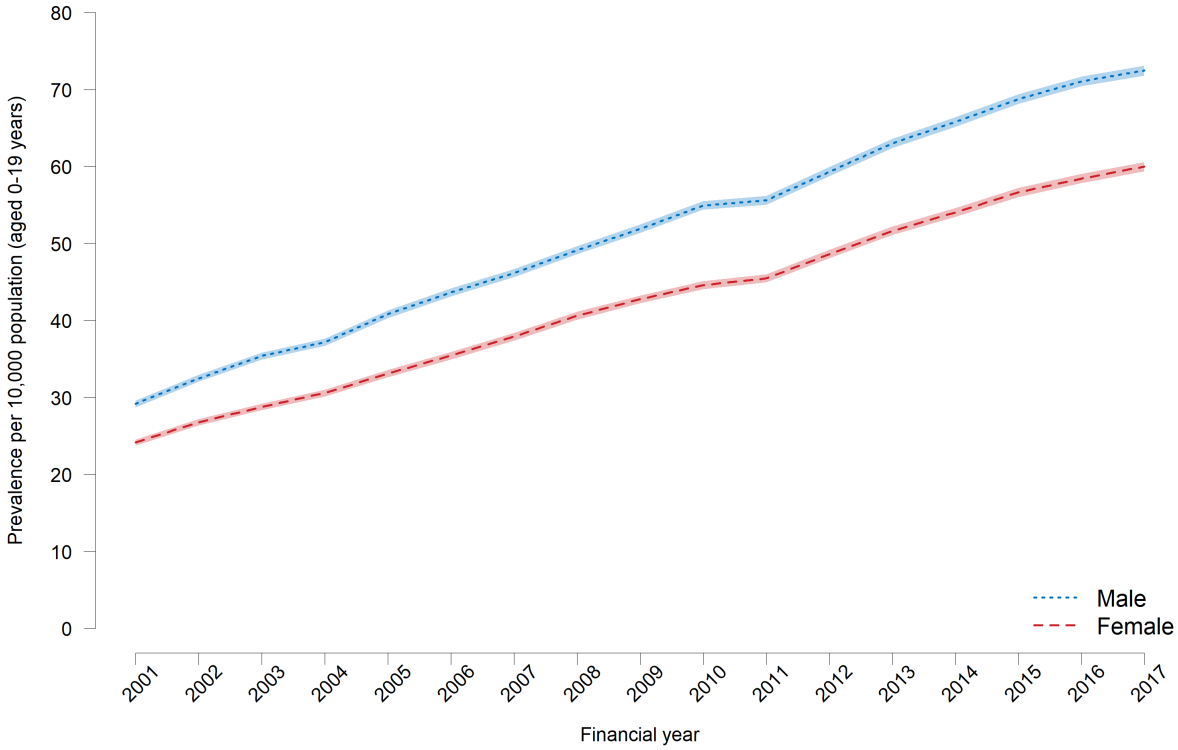


***Figure 7: Prevalence of life-limiting conditions (with 95% confidence intervals in lighter shading) in children (age 0-19) by sex for 2001/02-2017/18***

The prevalence of LLCs was significantly higher among boys (72.5 per 10,000 [95%CI 71.8-73.1]) than girls (60.0 per 10,000 [95%CI 59.4-60.6]) during 2017/18, though the increase in prevalence was similar for both sexes during the study time period (Figure 7).

Due to poor recording of ethnicity during 2001/02 and 2002/03 (7 & 6% missing data respectively), prevalence per ethnic group for these years was not calculated. Prevalence of LLCs was highest amongst children of Pakistani origin (103.9 per 10,000 [95%CI: 101.2-106.6]) (2017/18), and lowest among children of Chinese origin (32.0 per 10,000 [95% CI: 28.1-35.8] (2017/18)) (Figure 8). The rise in prevalence was similar between all ethnic groups.

Prevalence of LLCs was highest in the North West of England and Yorkshire and the Humber and lowest in the East Midlands (Figure 9). The increase in prevalence was similar in all Government Office Regions (see Supplementary Table 2 and Supplementary Table 3).

The prevalence of LLCs was highest in the most deprived group (88.6 per 10,000 [95% CI: 87.0-90.2] (2017/18)) and there was a gradient with deprivation with the lowest prevalence in the least deprived group (48.7 per 10,000 [95% CI: 47.5-49.9] (2017/18)) (Figure 10). This pattern was consistent over time.

The deprivation categories were population weighted therefore you would expect ~10% of children to have a LLC in each category. During the 17 year period, the proportion of children with a LLC in the least deprived area decreased from 8.1% to 7.3%. Changes in the proportion of children with an LLC in the other deprivation categories were smaller with the proportion of children with a LLC increasing in the five most deprived groups (Figure 10).

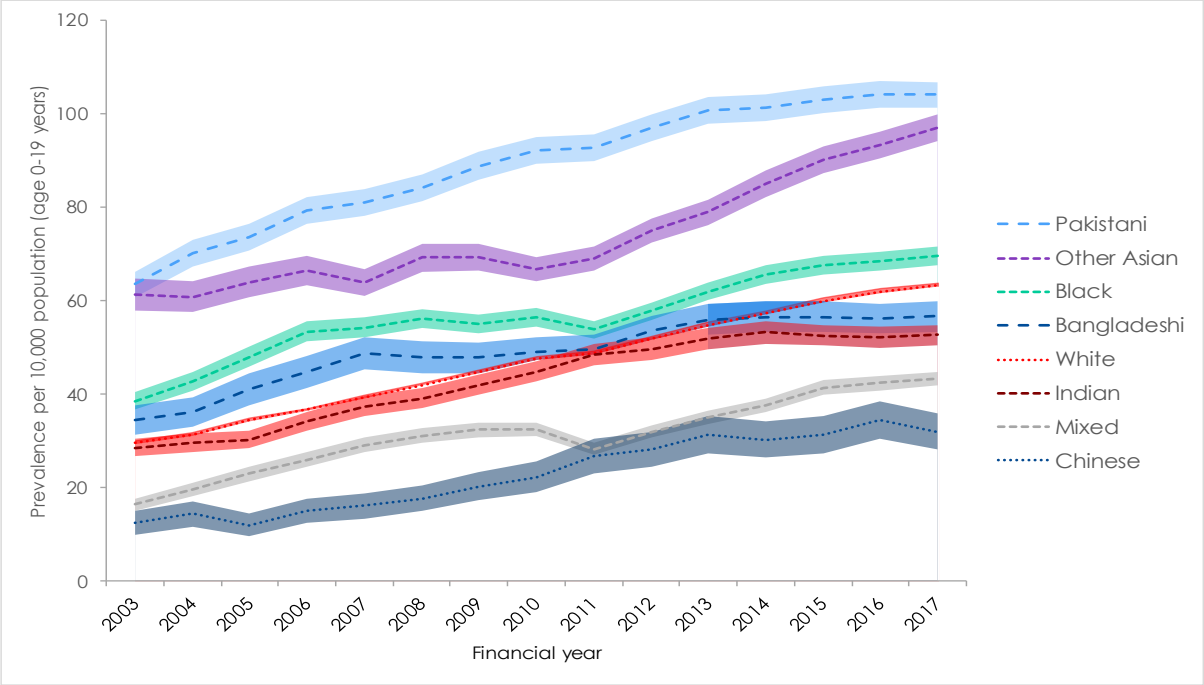


***Figure 8: Prevalence of life-limiting conditions (with 95% confidence intervals in lighter shading) in children (age 0-19) by ethnic group for 2001/02-2017/18***

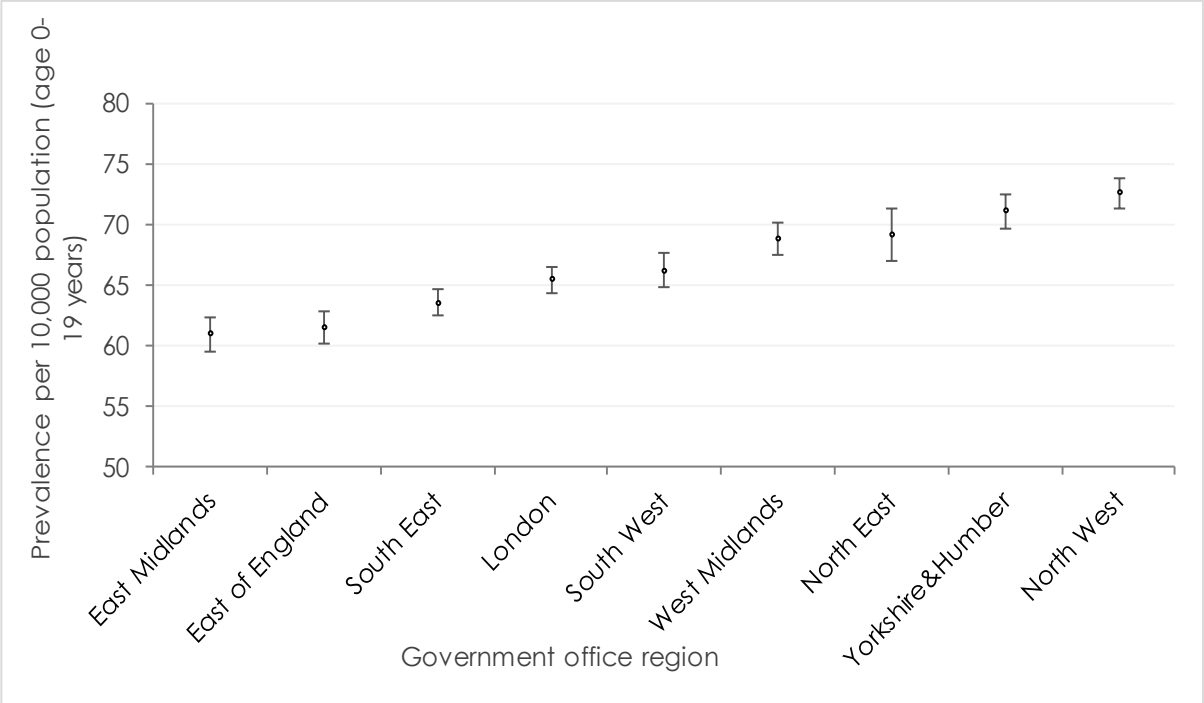


***Figure 9: Prevalence of life-limiting conditions in children by Government Office Region for 2017/18***

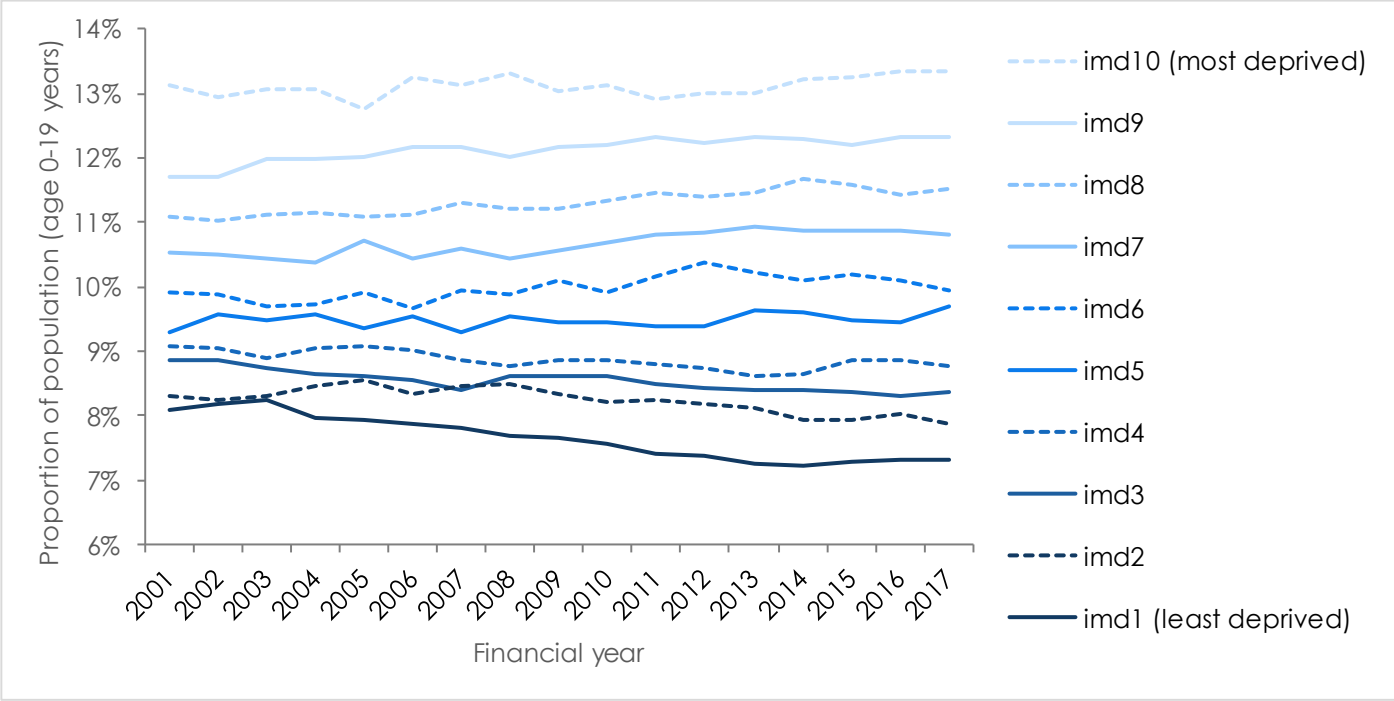


***Figure 10: Percentage of children (age 0-19) with life-limiting conditions by (population weighted) deprivation group for 2001/02-2017/18***

## Sensitivity analysis

In order to assess the impact of excluding certain ICD-10 codes from the definition of a LLC a sensitivity analysis was carried out with the following results (Supplementary Table 4):

(i) Exclusion of perinatal disorders after the first birthday made a small impact on the prevalence, mostly in the later years, reducing the overall prevalence in 2017/18 from 66.4 per 10,000 [95%CI 66.0-66.8] to 63.2 per 10,000 [95%CI 62.8-63.6].

(ii) The exclusion of oncology cases 5 years after diagnosis also made a small impact on the prevalence, reducing it by 2.3 per 10,000 to 64.1 per 10,000 [95%CI 63.6-64.5] in 2017/18.

(iii) Removal of non-central nervous system (CNS) oncology cases 5 years after diagnosis had a similar effect as the removal of all oncological cases.

(iv) Removal of early stage (1-3) kidney disease, made no discernible impact on the overall prevalence.

Combining all of the restricted definitions reduced the prevalence from 66.4 to 61.1 per 10,000 (95%CI 60.7-61.5) in 2017/18 (Figure 11).

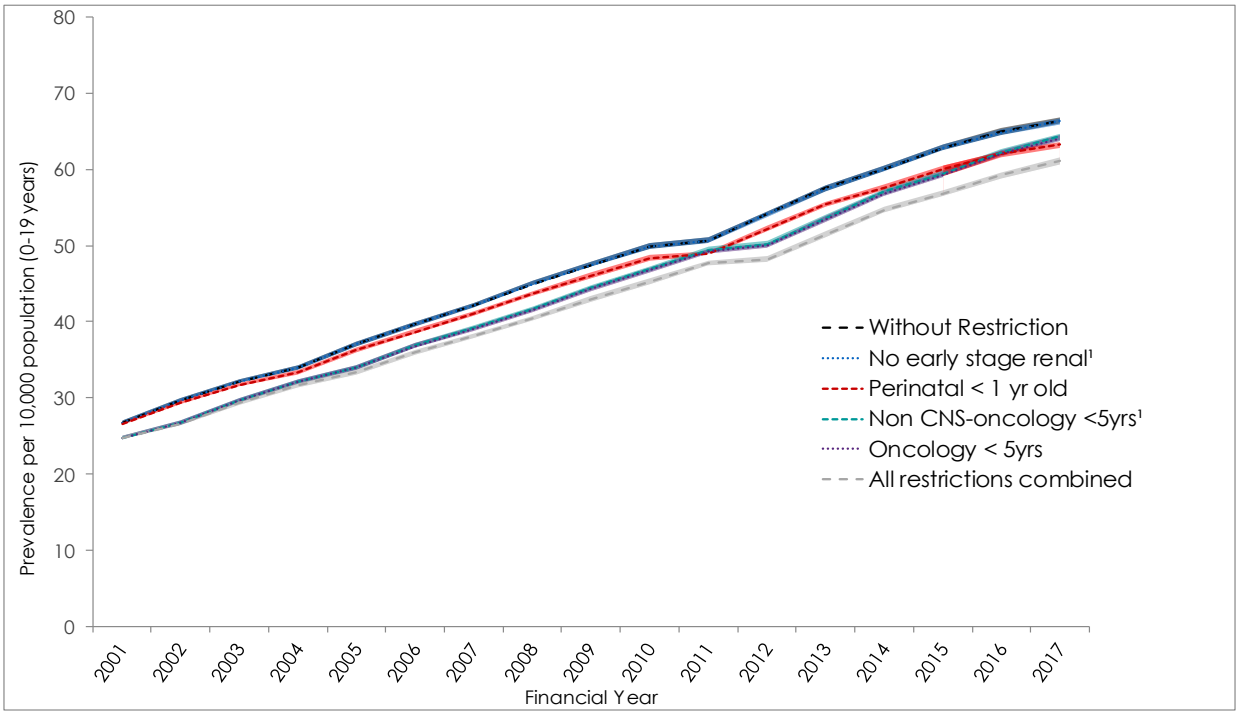


***Figure 11: Prevalence of life-limiting conditions (with 95% confidence intervals in lighter shading) in children (age 0-19) with restricted definitions of life-limiting condition.***
***[1]Lines overlap those of group above***

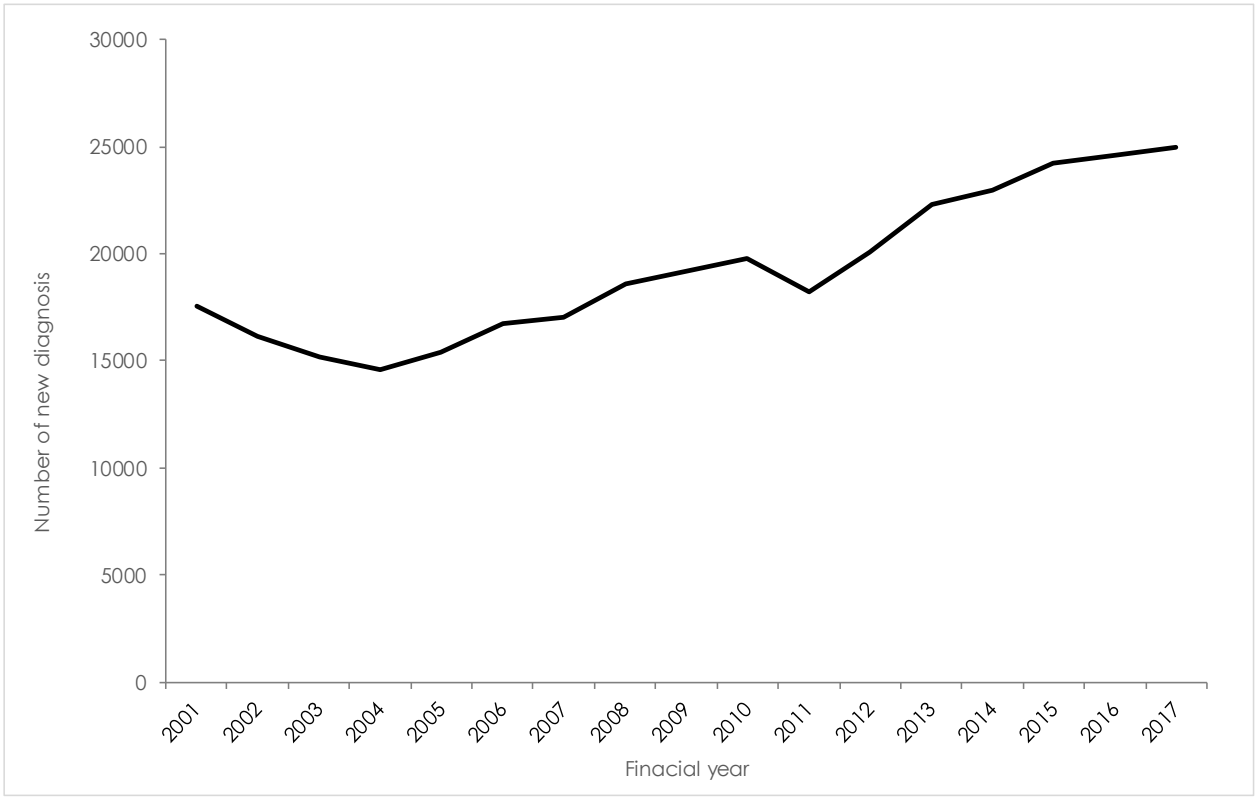


***Figure 12: Number of new diagnoses per year for children (age 0-19) for 2000/01-2017/18***

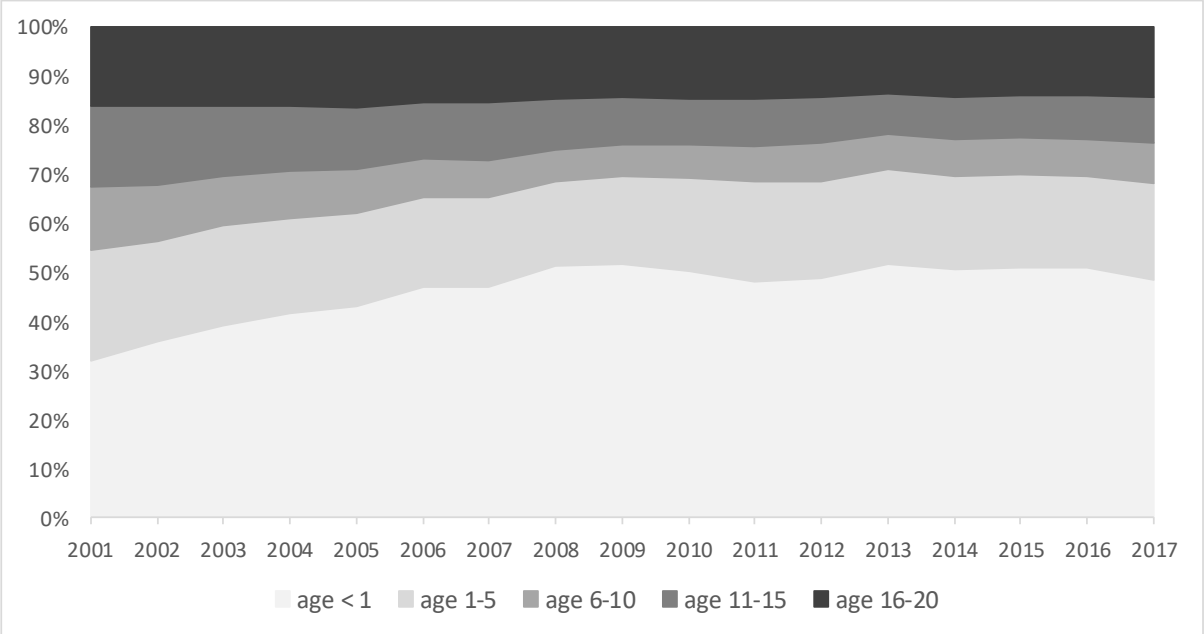


***Figure 13: Proportion of new diagnoses for children (age 0-19) with life-limiting conditions by age group for 2000/01-2017/18***

## Assessment of change in prevalence

### Change in incidence

The number of new recorded diagnoses increased steadily between 2001/2-2017/18 (Figure 12), with the majority of diagnoses (44%) occurring before the first birthday, and fewer diagnoses being made as age increased. The age at which a diagnosis was recorded changed over time with a higher proportion of diagnoses being made in the <1 year group in 2017/18 (48%) than in 2001/02 (32%) (Figure 13). However, for the earlier years, this could be due to left censoring effects i.e. individuals had been previously diagnosed before the start of this dataset (2000/01) (Figure 13). After exclusion of <1 year olds, the median age at which LLCs were diagnosed was 9 years 7 months. Over the 17 year period the median age of diagnosis (including under 1s) declined from 4 years 7 months to 1 year 1 months (Supplementary figure 2), although again, for the first few years this could be partly due to left edge effects.

The mean and median numbers of hospital admissions per child (as measured by continuous inpatient spells), remained fairly constant ranging from an average of 4.6 admissions per child in 2001/02 to 3.6 in 2017/18.

### Change in survival

Of the 359,643 individuals in the dataset, 10.4% (n=37,328) died during the study period 8.4% (n=30,187) of whom died before age 20. The proportion of children that died each year fell from 6.6% in 2001 to 2.3% in 2017 (Figure 14).

The median age of death (among 0-19 years) was between 0-1 years, or 1-2 years when including deaths beyond the cohort age (Figure 15). After exclusion of <1 year olds, which represent around 45% of all deaths among children with LLCs, the median age of death increased to around age 12 (for death at any age) (Figure 15). As a proportion of children who died the majority of death occurred among the congenital group (27.2-36%) (Table 3). The

proportion of deaths for perinatal disorders rose from 15.6% to 31.4% of all deaths whilst the proportion of deaths among oncology cases declined.

In addition to the 30,187 deaths between 2001/02-2017/18, a further 7,605 deaths were recorded for individuals who had a LLC but were over the age of 20 (Table 4). The number increased steadily, although dropped in 2017 which is likely to be due to a lag in reporting deaths.

Restricting the analysis of death to those who were born within the time period of the study (Supplementary Table 5), showed that 17% of those born in 2001, and subsequently diagnosed with a LLC, died during the time period of this study. This reduced in the latter years, partly because these children are all still young and partly given the lag in death certification that may have occurred.

Finally, using a cohort of those only born after the study period (31st March 2001), the Kaplan-Meier survival analysis showed that the majority of deaths occurred within the first year of diagnosis and that the survival to age 17 for those born in 2001 was 88% (Figure 16).

These analyses suggest that both change in incidence or recording of diagnoses and an increase in survival may be contributing to the overall increase in prevalence in this population.

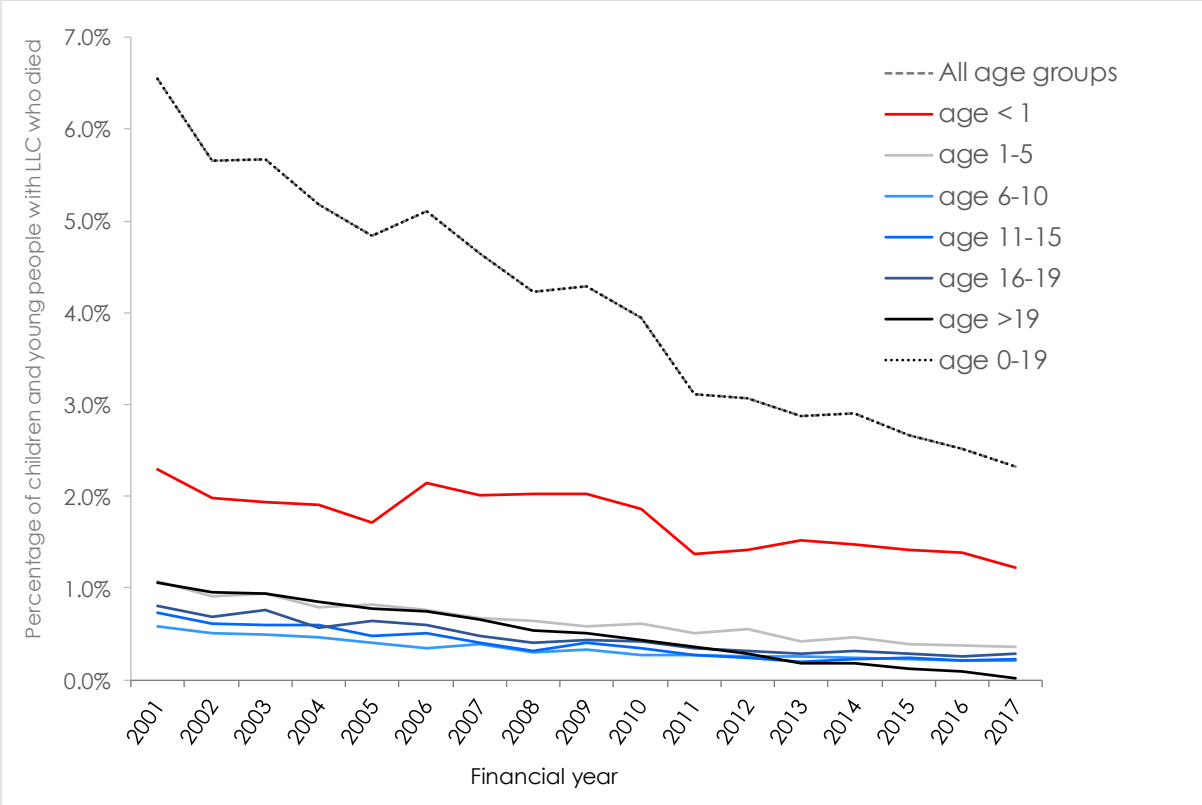


***Figure 14: Proportion of children (age 0-19) with a life-limiting condition who died per year***

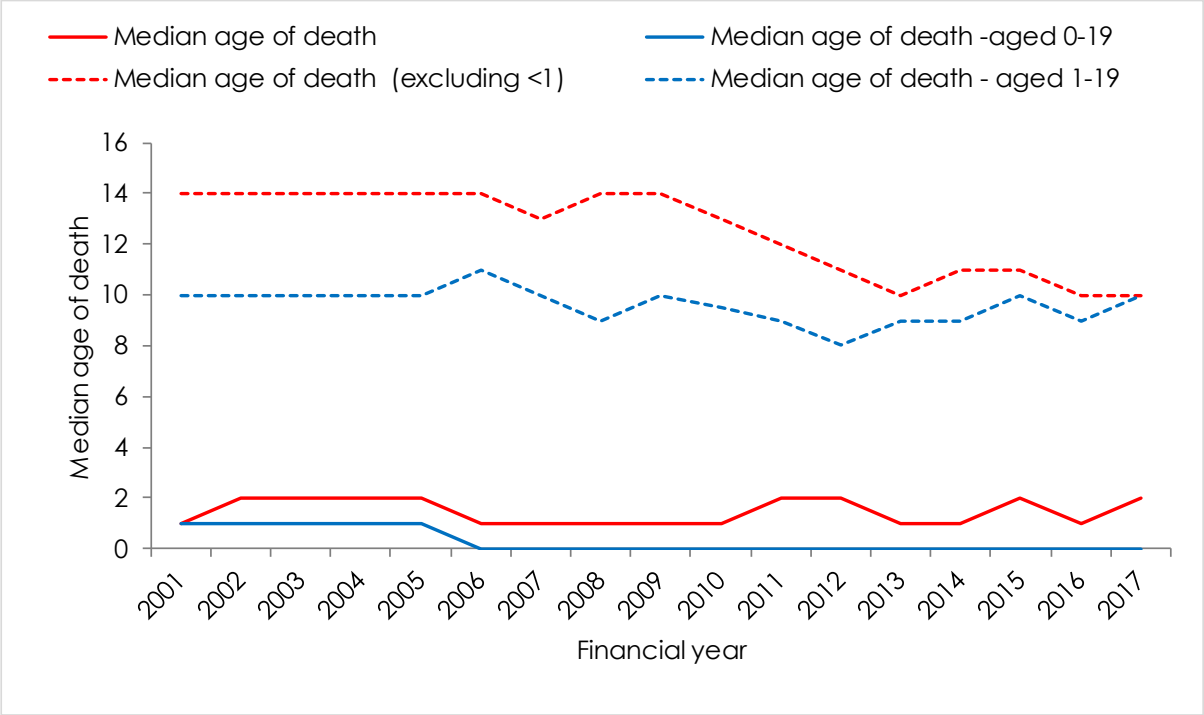


***Figure 15: Median age of death of children with a life-limiting condition (2000/01-2017/18)***

Table 3: Percentage of children (age 0-19) who died by diagnostic group

| Main diagnostic group | Number of deaths | | % deaths (as a percentage of all deaths) | Total cohort | % (as % of total) |
|---|---|---|---|---|---|
| | Aged 0-19 | Aged >19 | | | |
| **Circulatory** | 966 | 271 | 3.3% | 8,575 | 11% |
| **Congenital** | 8,484 | 1,403 | 26.5% | 122,741 | 7% |
| **Gastrointestinal** | 392 | 92 | 1.3% | 4,609 | 9% |
| **Genitourinary** | 1,242 | 354 | 4.3% | 20,107 | 6% |
| **Haematology** | 531 | 198 | 2.0% | 18,216 | 3% |
| **Metabolic** | 1,124 | 365 | 4.0% | 12,998 | 9% |
| **Neurology** | 3,679 | 1,688 | 14.4% | 38,622 | 10% |
| **Oncology** | 5,498 | 1,531 | 18.8% | 47,399 | 12% |
| **Other** | 349 | 91 | 1.2% | 4,845 | 7% |
| **Perinatal** | 6,473 | 293 | 18.1% | 51,439 | 13% |
| **Respiratory** | 1,449 | 855 | 6.2% | 30,083 | 5% |
| **Total** | 30,187 | 7141 | | 359,634 | |

| Table 4: Number of individuals that died after age 20 by financial year | | |
|---|---|---|
| **Financial year** | **Number who died aged 0-19** | **Number who died aged > 19** |
| **2001** | 1,660 | 483 |
| **2002** | 1,540 | 510 |
| **2003** | 1,676 | 545 |
| **2004** | 1,606 | 529 |
| **2005** | 1,638 | 530 |
| **2006** | 1,910 | 552 |
| **2007** | 1,862 | 527 |
| **2008** | 1,885 | 454 |
| **2009** | 2,048 | 473 |
| **2010** | 1,973 | 480 |
| **2011** | 1,537 | 414 |
| **2012** | 1,704 | 349 |
| **2013** | 1,771 | 295 |
| **2014** | 1,871 | 319 |
| **2015** | 1,875 | 251 |
| **2016** | 1,842 | 238 |
| **2017** | 1,789 | 192 |
| **Total** | 30,187 | 7,141 |

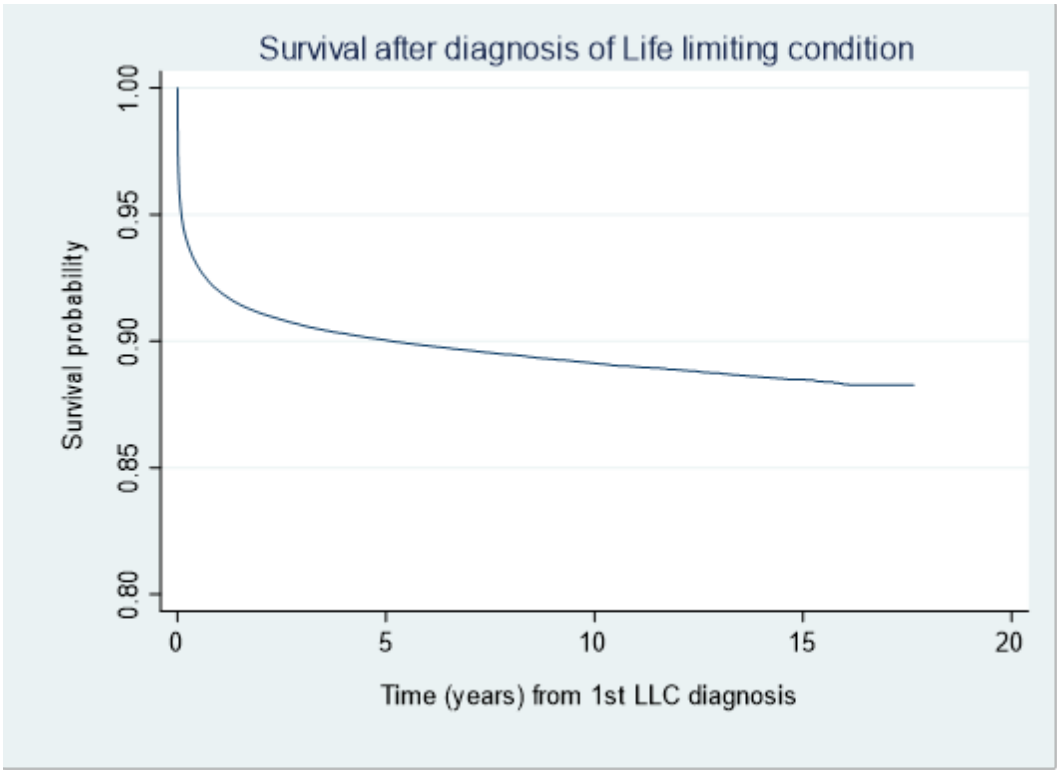


***Figure 16: Kaplan-Meier survival curve for the cohort of children born after 31st March 2001***

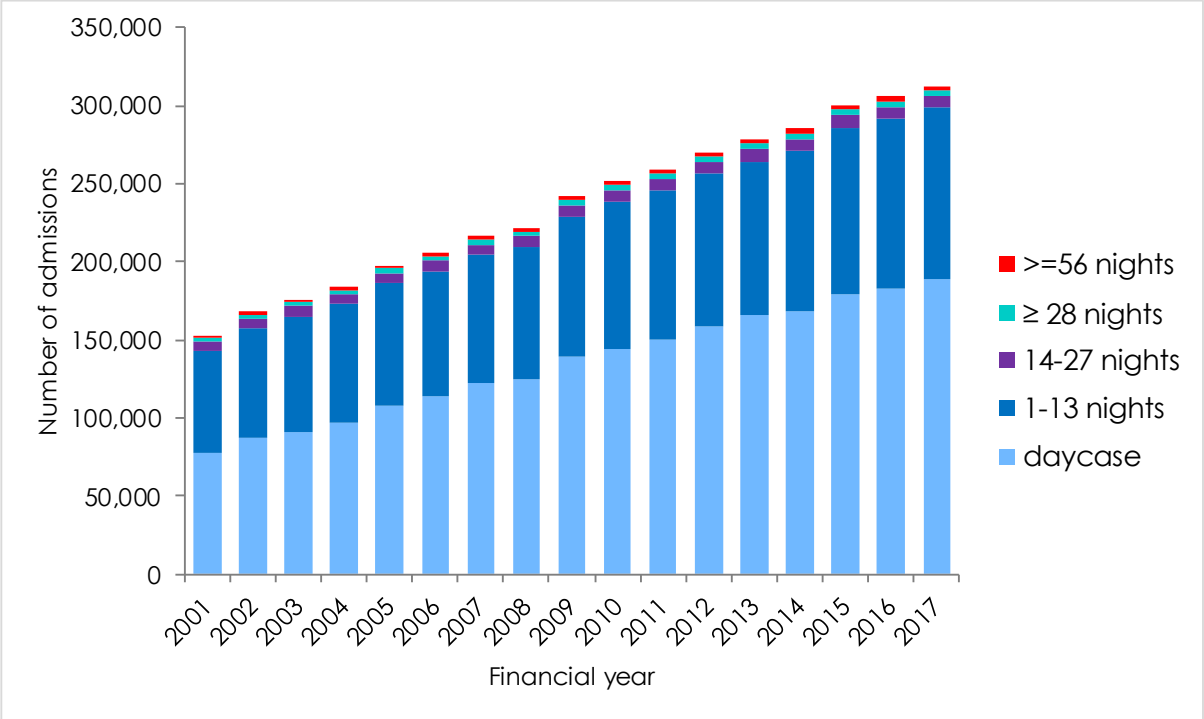


***Figure 17: Number of hospital admissions for children (aged 0-19) with life-limiting conditions***

## Additional Analyses

### Proxy of Complexity

Although the mean and median length of stay remained constant during the 17 year period, the total number of admissions increased. However, this was driven by a large increase in the number of day cases (from 51 to 61% of all admission) (Figure 17, Supplementary Table 7).

Supplementary Table ***7*** shows that the absolute number of hospital admissions lasting 28 days or more increased between 2001/02-2017/18. Although the absolute number of hospital admissions increased, the proportions of individuals with a maximum length of stay of 28 nights or more remained constant at 1.9-2.5% of all admissions and 4.9-5.5% of all admissions with an overnight stay. The proportion of emergency admittance also remained constant for those with a stay of 28 days or more and the proportion of emergency admittance increased for those with a stay between 1-13 nights.

Factors that increased the odds of having a longer stay in hospital (compared to hospital stays <28 days) were; being under age 1 year (compared to being age 1-5 years) and having a non-emergency admission (Table 5). Compared to having a congenital disorder, having almost any other diagnosis increased the odds of having a hospital stay of ≥28 days, with oncological cases having the greatest risk (OR 3.08 [95%CI 2.99-3.19]). These results remained unchanged when birth admissions were excluded, with the exception of having a stay of at least 28 days, which was less likely to be associated with a non-emergency admission (Supplementary Table 8).

### Number of young people who would require adult services each year

As the number of children who died each year at age 19 remained relatively static, the numbers surviving into adulthood increased in line with the increasing prevalence of LLCs (Supplementary Table 9).

Table 5: Hierarchical Logistic regression model (dependent variable Length of stay ≥ 28 days)

| | Odds Ratio | SE | Z | P>[z] | 95% Confidence Interval | |
|---|---|---|---|---|---|---|
| Year | 0.97 | 0.001 | -22.47 | <0.01 | 0.97 | 0.98 |
| **Sex** | | | | | | |
| Male | 1.00 (reference) | | | | | |
| Female | 1.05 | 0.010 | 5.5 | <0.01 | 1.03 | 1.07 |
| **Age group** | | | | | | |
| age < 1 | 11.39 | 0.013 | 185.54 | <0.01 | 11.10 | 11.68 |
| age 1-5 | 1.00 (reference) | | | | | |
| age 6-10 | 0.71 | 0.019 | -18.34 | <0.01 | 0.68 | 0.73 |
| age 11-15 | 0.94 | 0.018 | -3.65 | <0.01 | 0.90 | 0.97 |
| age 16-19 | 1.24 | 0.018 | 12.1 | <0.01 | 1.20 | 1.28 |
| **Ethnic group** | | | | | | |
| White | 1.00 (reference) | | | | | |
| Missing | 0.71 | 0.031 | -11.22 | <0.01 | 0.67 | 0.75 |
| Indian | 1.11 | 0.030 | 3.49 | <0.01 | 1.05 | 1.18 |
| Pakistani | 1.22 | 0.020 | 9.61 | <0.01 | 1.17 | 1.27 |
| Bangladeshi | 1.31 | 0.039 | 6.85 | <0.01 | 1.21 | 1.41 |
| Black | 1.41 | 0.021 | 16.17 | <0.01 | 1.35 | 1.47 |
| Chinese | 1.31 | 0.079 | 3.43 | <0.01 | 1.12 | 1.53 |
| Mixed | 1.14 | 0.026 | 4.8 | <0.01 | 1.08 | 1.20 |
| Other Asian | 1.38 | 0.022 | 14.63 | <0.01 | 1.32 | 1.45 |
| **GOR** | | | | | | |
| London | 1.00 (reference) | | | | | |
| East Midlands | 1.07 | 0.021 | 3.1 | <0.01 | 1.02 | 1.11 |
| East of England | 0.98 | 0.020 | -1.28 | 0.20 | 0.94 | 1.01 |
| North East | 0.94 | 0.026 | -2.55 | 0.01 | 0.89 | 0.98 |
| North West | 1.07 | 0.018 | 3.94 | <0.01 | 1.04 | 1.11 |
| South East | 0.97 | 0.018 | -1.64 | 0.10 | 0.94 | 1.01 |
| South West | 1.04 | 0.020 | 1.89 | 0.06 | 1.00 | 1.08 |
| West Midlands | 1.02 | 0.019 | 1.1 | 0.27 | 0.98 | 1.06 |
| Yorkshire & Humber | 1.01 | 0.020 | 0.44 | 0.66 | 0.97 | 1.05 |
| **Deprivation** | | | | | | |
| imd1 (least deprived) | 0.79 | 0.017 | -13.94 | <0.01 | 0.77 | 0.82 |
| imd2 | 0.84 | 0.016 | -10.86 | <0.01 | 0.82 | 0.87 |
| imd3 | 0.87 | 0.015 | -9.82 | <0.01 | 0.84 | 0.89 |
| imd4 | 0.92 | 0.013 | -5.94 | <0.01 | 0.90 | 0.95 |
| imd 5 (most deprived) | 1.00 (reference) | | | | | |
| **Diagnoses** | | | | | | |

| | | | | | | |
|---|---|---|---|---|---|---|
| Congenital | 1.00 (reference) | | | | | |
| Circulatory | 1.65 | 0.031 | 16.27 | <0.01 | 1.55 | 1.75 |
| Gastrointestinal | 3.05 | 0.042 | 26.56 | <0.01 | 2.81 | 3.31 |
| Genitourinary | 2.18 | 0.024 | 31.94 | <0.01 | 2.08 | 2.29 |
| Haematology | 1.49 | 0.025 | 16.26 | <0.01 | 1.42 | 1.56 |
| Metabolic | 1.12 | 0.029 | 3.85 | <0.01 | 1.06 | 1.19 |
| Neurology | 1.74 | 0.018 | 31.53 | <0.01 | 1.68 | 1.80 |
| Oncology | 3.11 | 0.017 | 68.07 | <0.01 | 3.01 | 3.21 |
| Other | 2.44 | 0.036 | 24.52 | <0.01 | 2.27 | 2.62 |
| Perinatal | 0.82 | 0.015 | -12.47 | <0.01 | 0.80 | 0.85 |
| Respiratory | 2.02 | 0.017 | 42.14 | <0.01 | 1.96 | 2.09 |
| **Emergency Admission** | | | | | | |
| Non-emergency emission | 1.00 (reference) | | | | | |
| Emergency | 0.87 | 0.01 | -14.47 | <0.01 | 0.86 | 0.89 |

## Modelling of future prevalence

### Estimated Future Prevalence in England

The most conservative estimates (**Model 3**) predicted that the number of children with LLCs would rise from 87,572 (95% CI 88,462-86,726) in 2017 to 96, 275 (95%CI 95,318-97,242) in 2030 (Figure 18) equating to a change prevalence from 67.1 (95%CI 67.8-66.5) to 67.0 (95%CI 67.7-66.3) per 10,000 ( Figure 19).

Less conservative estimates (**Model 2**) using the restricted definition of LLCs, predicted that the number of children with a LLC would rise from 81,265 (95%CI 82,259-80,284) in 2017 to 116,770 (95%CI 108,894-125,209) in 2030 equating to a prevalence of 81.26 (95%CI 75.78-87.13) per 10,000 in 2030.

The least conservative estimates (**Model 1**), which used the broadest definition of LLCs, predicted that the number of children with LLCs would rise to 121,023 (95%CI 113,031-129,573) by 2030 or a prevalence of 84.22 (95%CI 78.66-90.17) per 10,000.

Stratification by GOR, indicated that the rise in numbers would be greatest in London followed by the South East and North West ( Figure 20), although due to differences in the estimation of population growth in the regions, the prevalence per 10,000 would be greatest in the North West at 92.37 (95%CI 82.23-103.74) ( Figure 21).

Modelling the probability of having a specific diagnosis predicted the largest increase in absolute numbers to be among those with a congenital disorder, estimated to affect 53,425 [95% CI 58,254-48,981] individuals by 2030 ( Figure 22). Likewise, the predicted prevalence was the largest for congenital disorders at 37.2 per 10,000 population [95%CI 40.5-34.1] ( Figure 23). Although the predicted prevalence increased for most diagnoses, the predicted prevalence remained static for oncology and circulatory disorders.

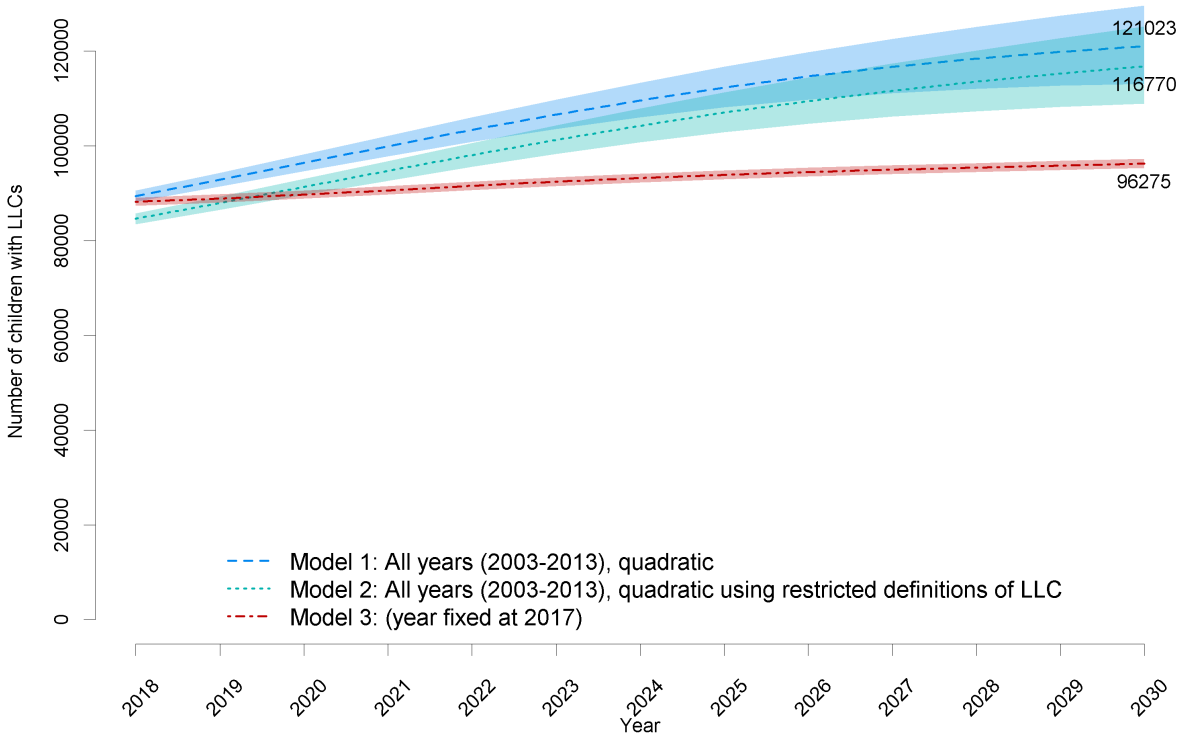


***Figure 18: Predicted numbers (with 95% confidence intervals in lighter shading) of children (age 0-19) with life-limiting conditions in England***

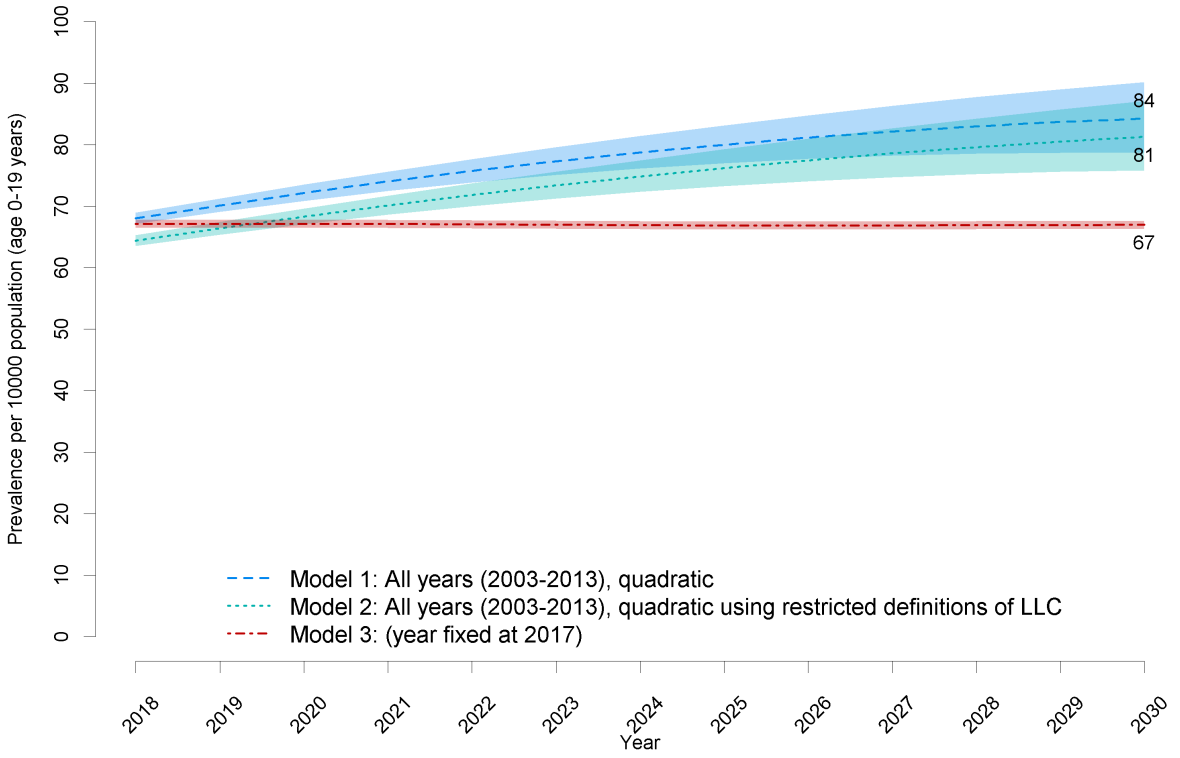


***Figure 19: Predicted prevalence (with 95% confidence intervals in lighter shading) of children (age 0-19) with life-limiting conditions in England***

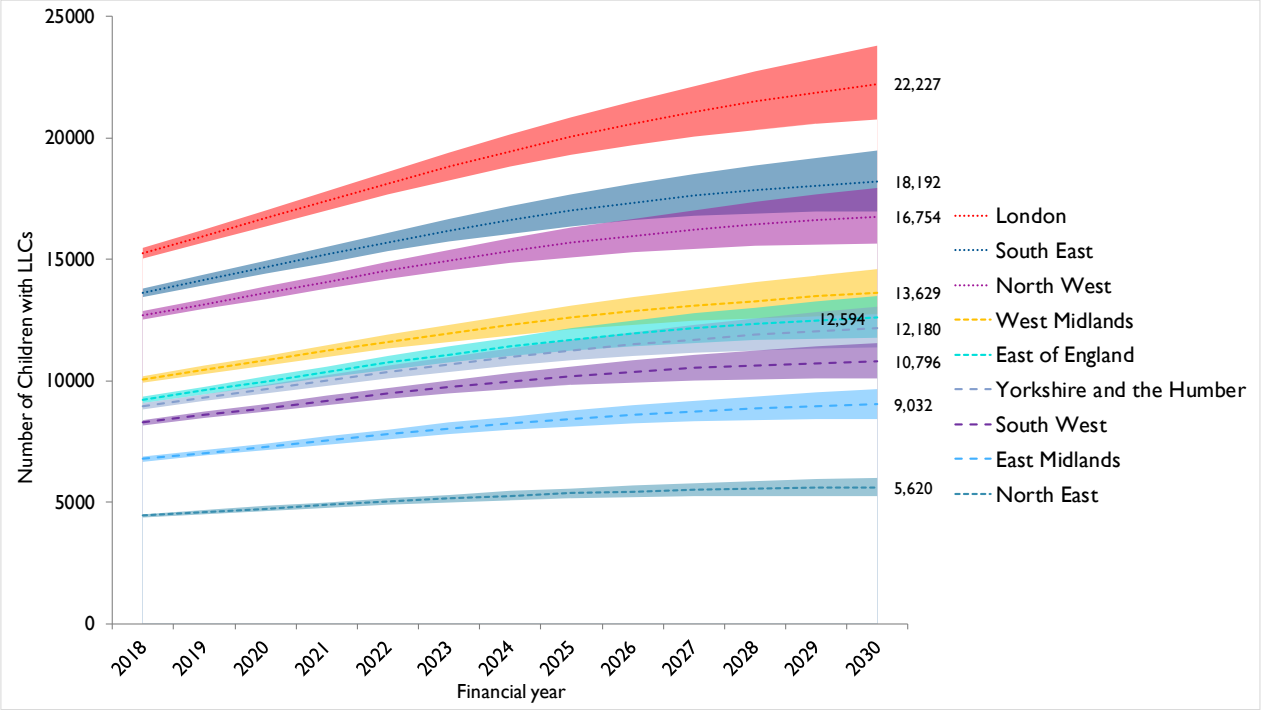


***Figure 20: Predicted number (with 95% confidence intervals in lighter shading) of children (age 0-19) with life-limiting conditions in England by Government Office Region***

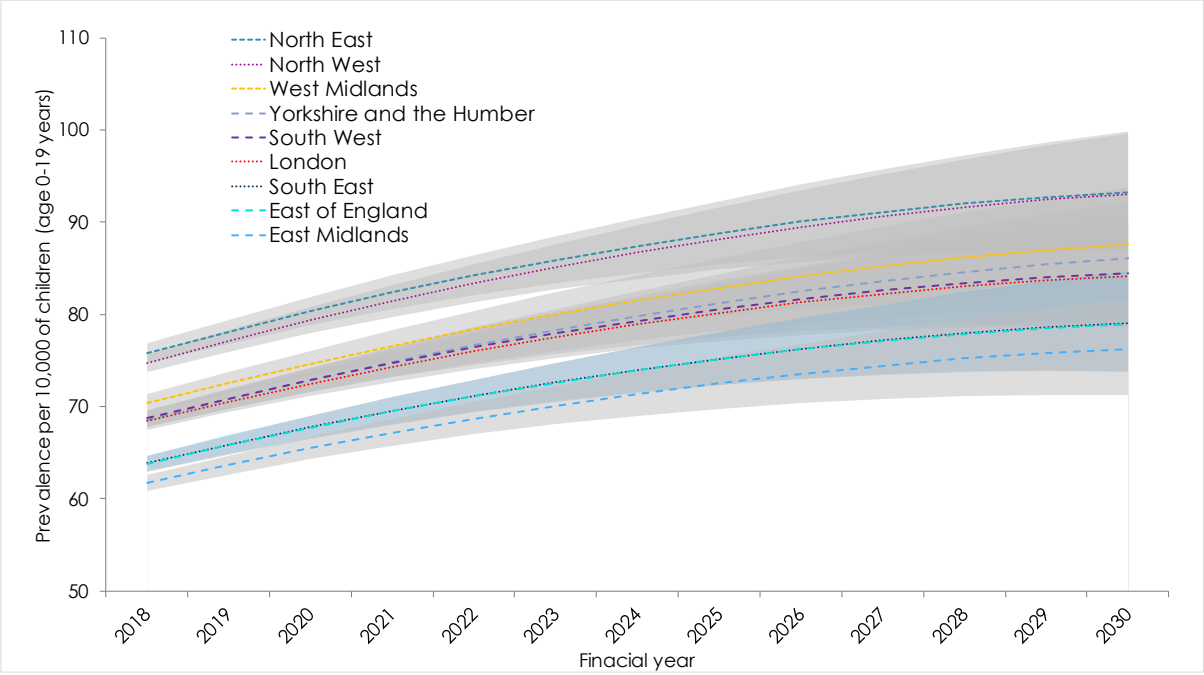


***Figure 21: Predicted prevalence of life-limiting conditions (with 95% confidence intervals in grey shading) per 10,000 of children (age 0-19) in England by Government Office Region***

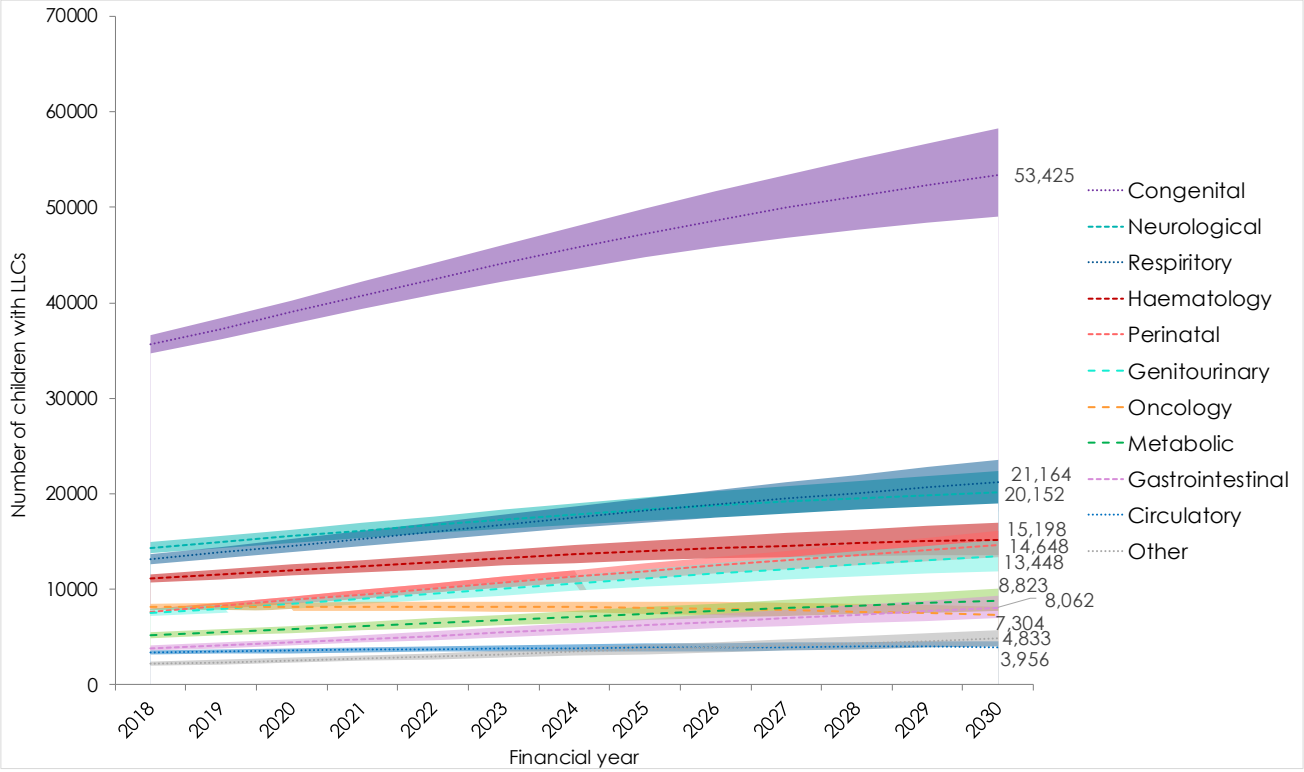


***Figure 22: Predicted numbers (with 95% confidence intervals in lighter shading) of children (age 0-19) with a life-limiting condition in England by diagnostic group***

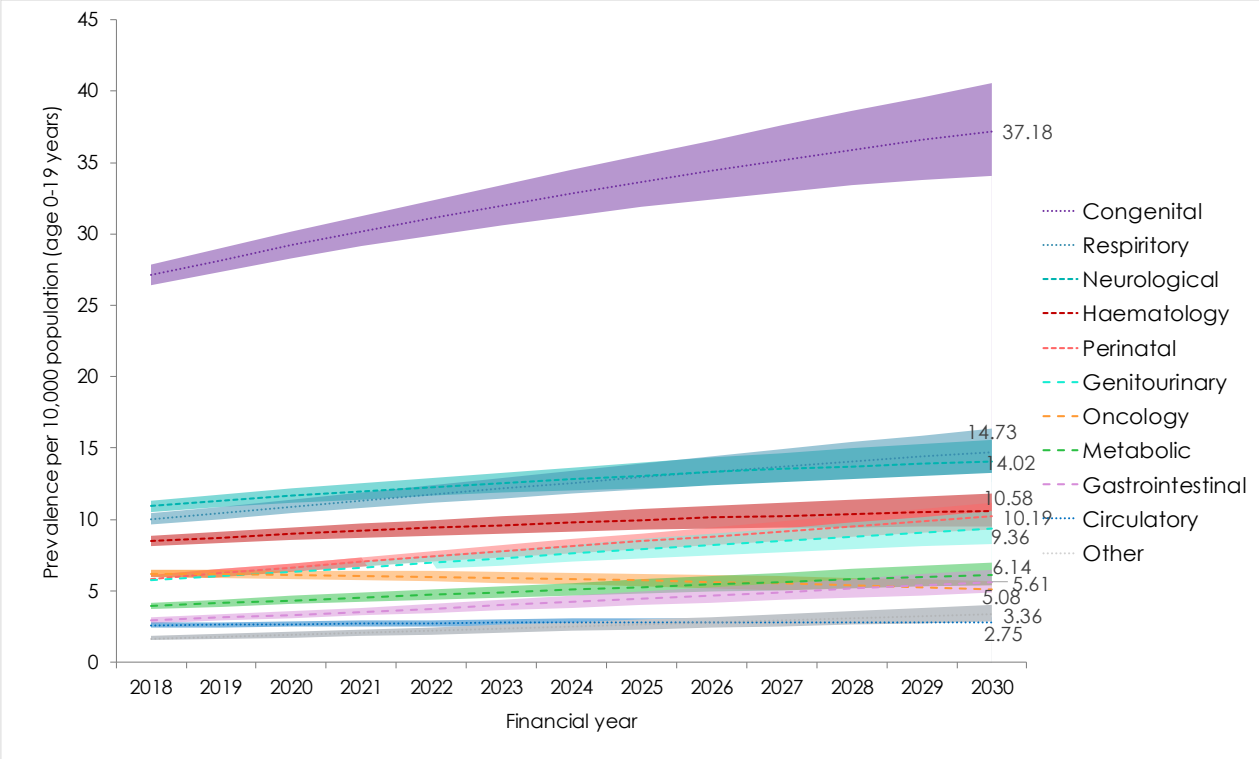


***Figure 23: Predicted prevalence of each specific diagnostic group per 10,000 population (with 95% confidence intervals in lighter shading) of children (age 0-19) in England***

### Estimated Future prevalence in Scotland, Wales and Northern Ireland

For Scotland ( Figure 24 & Figure 25), **Model 3** predicted an increase from 5933 (2018, 95%CI 5874-5993) to 6003 (2030, 95%CI 5943-6063), assuming no changes in incidence or survival since 2017. Using the restricted definition of LLCs (**Model 2**), but allowing year terms in the model, there was an increase from 5819 (95%CI 5761-5877) to 6449 (95%CI 6126-6771) individuals; under the broadest definition of LLCs (**Model 1**) there was an increase from 6051 (95%CI 5991-6112) to 6572 (95%CI 6243-6900) individuals. These translate to changes in prevalence from 51.35 (95%CI 50.84-51.87) to 50.99 (95%CI 50.48-51.50) per 10,000 (**Model 3** – no change), 50.36 (95%CI 49.86-50.87) to 54.78 (95%CI 52.04-57.52) per 10,000 (**Model 2**) and 52.38 to 55.83 per 10,000 (**Model 1**).

For Wales ( Figure 26 & Figure 27), **Model 3** predicted an increase from 3650 (2018, 95%CI 3614-3687) to 3711 (2030, 95%CI 3674-3748), assuming no changes in incidence or survival since 2017. Using the restricted definition of LLCs (**Model 2**), but allowing year terms in the model, there was an increase from 3580 (95%CI 3544-3616) to 3987 (95%CI 3788-4186) individuals; under the broadest definition of LLCs (**Model 1**) there was an increase from 3723 (95%CI 3686-3760) to 4063 (95%CI 3860-4266) individuals. These translate to changes in prevalence from 51.46 (95%CI 50.95-51.98) to 50.81 (95%CI 50.30-51.32) per 10,000 (**Model 3** - a decrease), 50.47 (95%CI 49.96-50.97) to 54.59 (95%CI 51.86-57.32) per 10,000 (**Model 2**) and 52.49 (95%CI 51.96-53.01) to 55.63 (95%CI 52.85-58.41) per 10,000 (**Model 1**).

For Northern Ireland ( Figure 28 & Figure 29), **Model 3** predicted an increase from 2497 (2018, 95%CI 2472-2522) to 2592 (2030, 95%CI 2566-2618), assuming no changes in incidence or survival since 2017. Using the restricted definition of LLCs (**Model 2**), but allowing year terms in the model, there is an increase from 2448 (95%CI 2424-2473) to 2784 (95%CI 2644-2923) individuals; under the broadest definition of LLCs (**Model 1**) there is an increase from 2546 (95%CI 2521-2572) to 2837 (95%CI 2695-2979) individuals. These translate to changes in prevalence from 51.93 (95%CI 51.44-52.47) to 51.57 (95%CI 51.05-52.08) per 10,000 (**Model 3** - a decrease), 50.92 (95%CI 50.41-51.43) to 55.38 (95%CI 52.61-58.15) per 10,000 (**Model 2**) and 52.96 (95%CI 52.43-53.49) to 56.45 (95%CI 53.63-59.28) per 10,000 (**Model 1**).

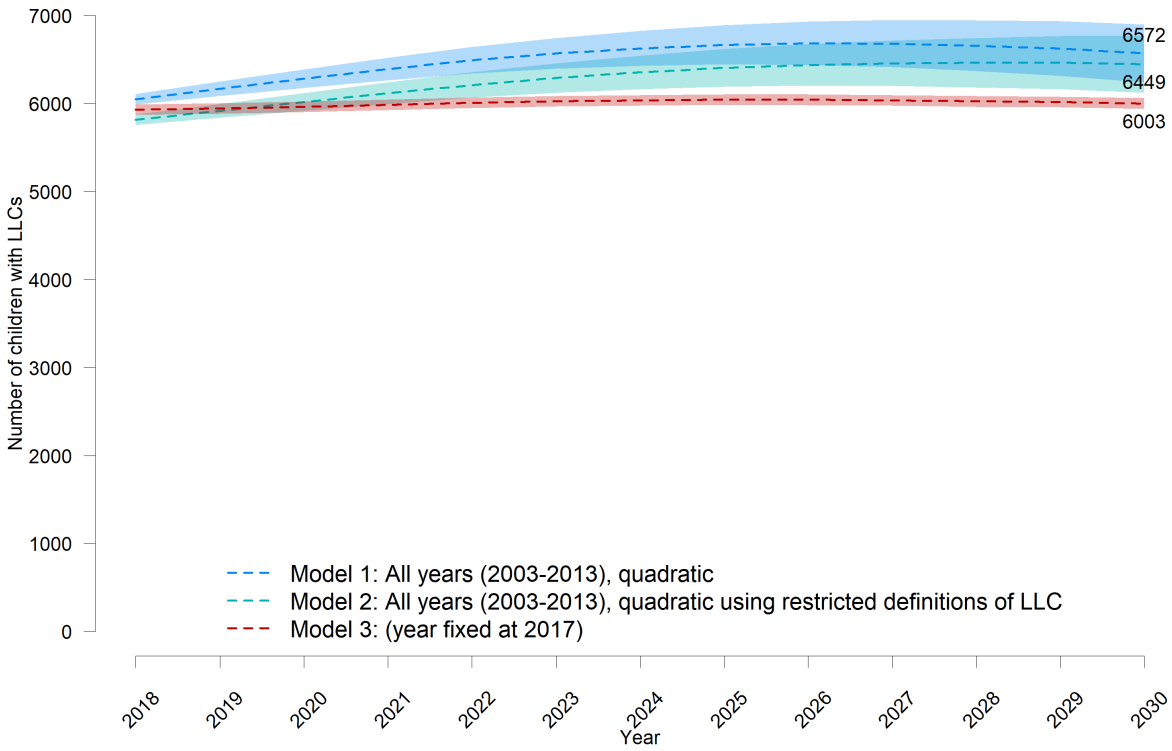


***Figure 24: Predicted number (with 95% confidence intervals in lighter shading) of children with a life-limiting condition (age 0-19) in Scotland***

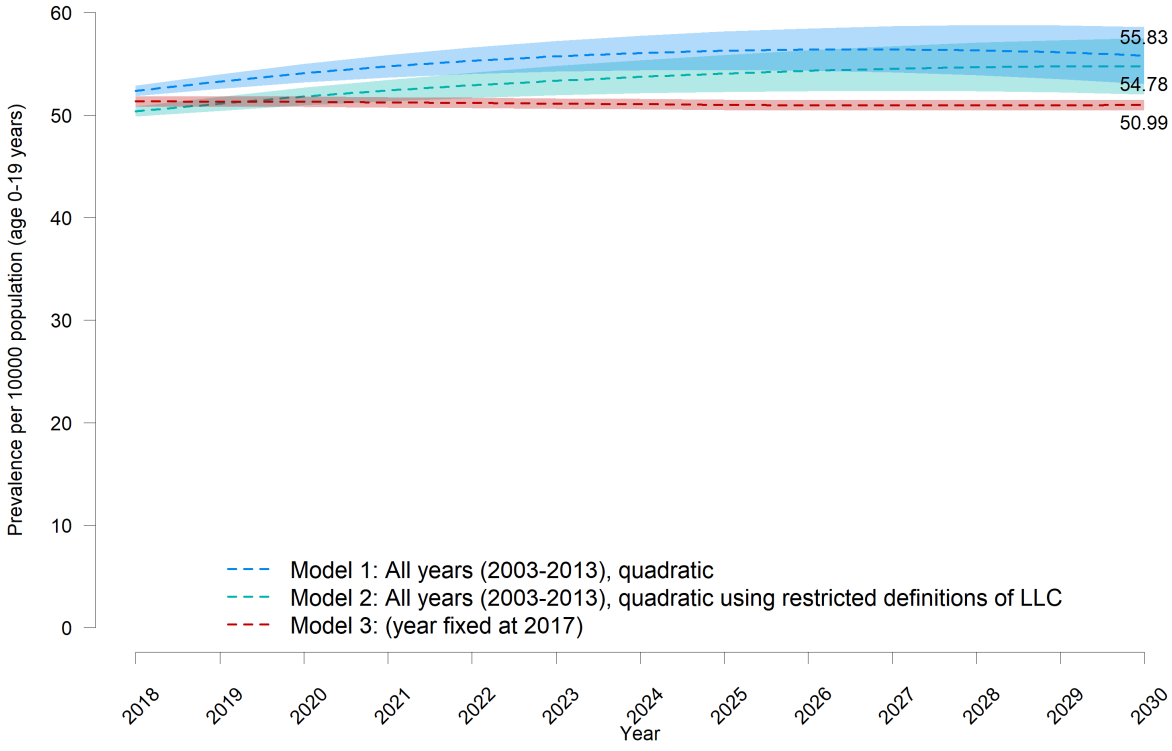


***Figure 25: Predicted prevalence (with 95% confidence intervals in lighter shading) of children with a life-limiting condition (age 0-19) in Scotland***

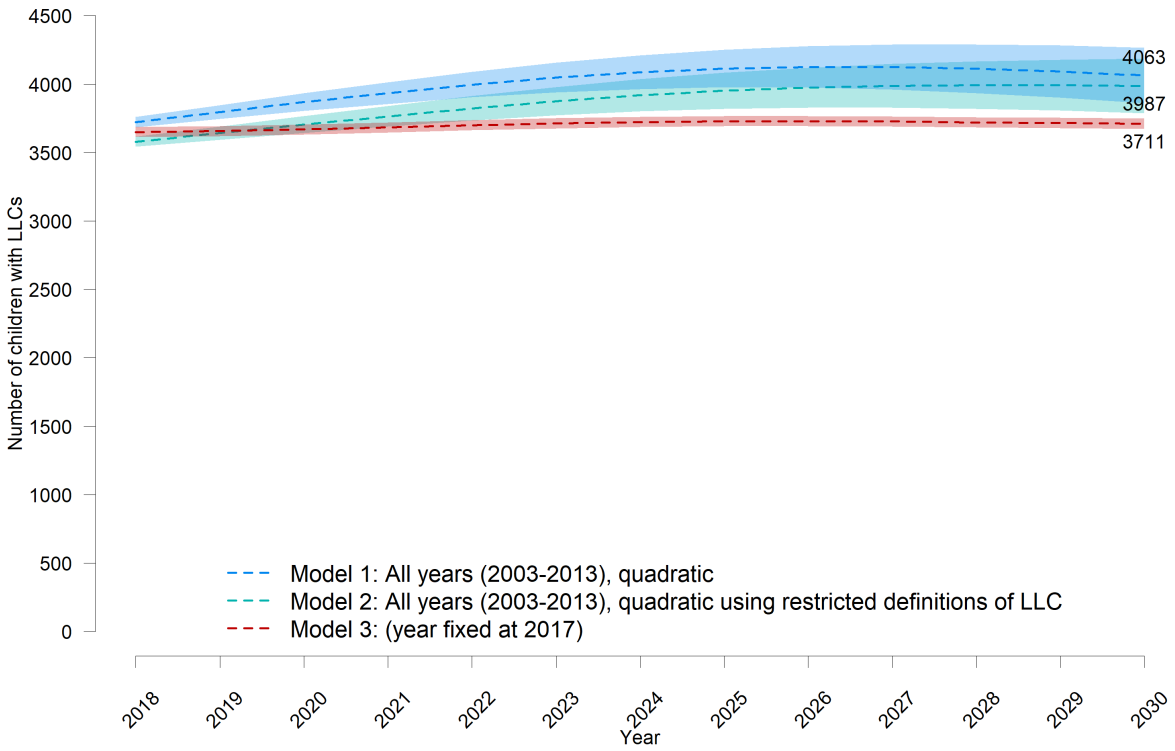


***Figure 26: Predicted number (with 95% confidence intervals in lighter shading) of children with a life-limiting condition (age 0-19) in Wales***

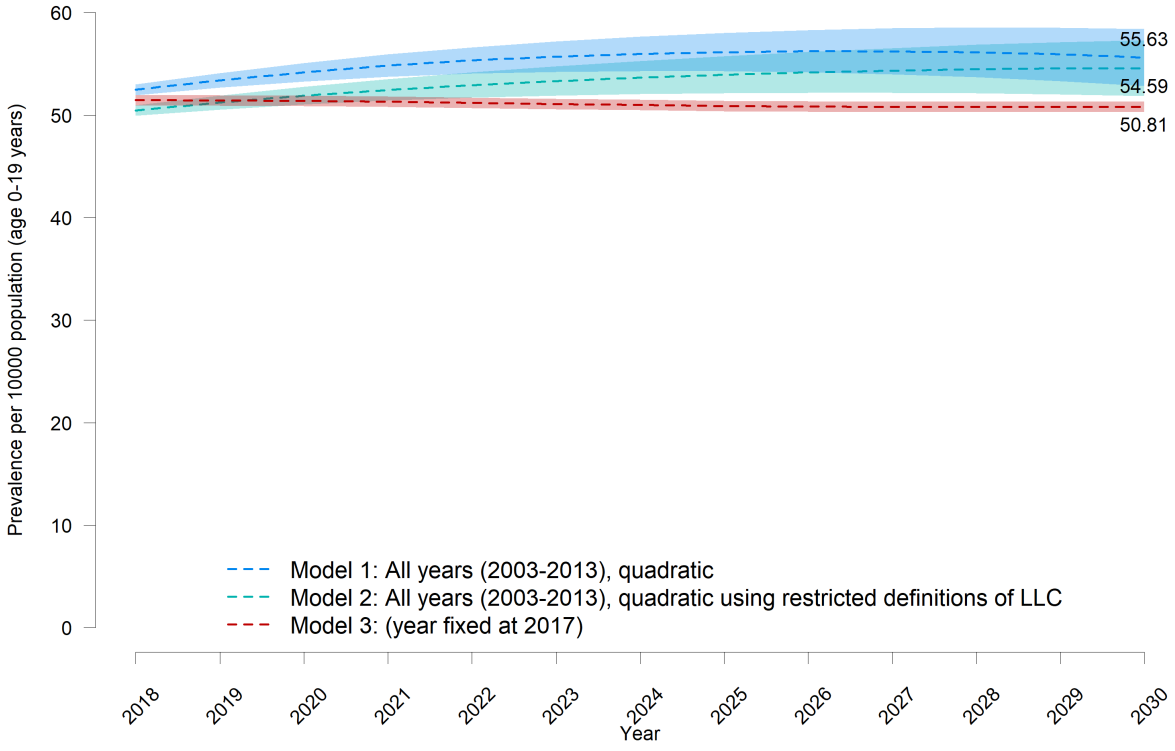


***Figure 27: Predicted prevalence (with 95% confidence intervals in lighter shading) of children with a life-limiting condition (age 0-19) in Wales***

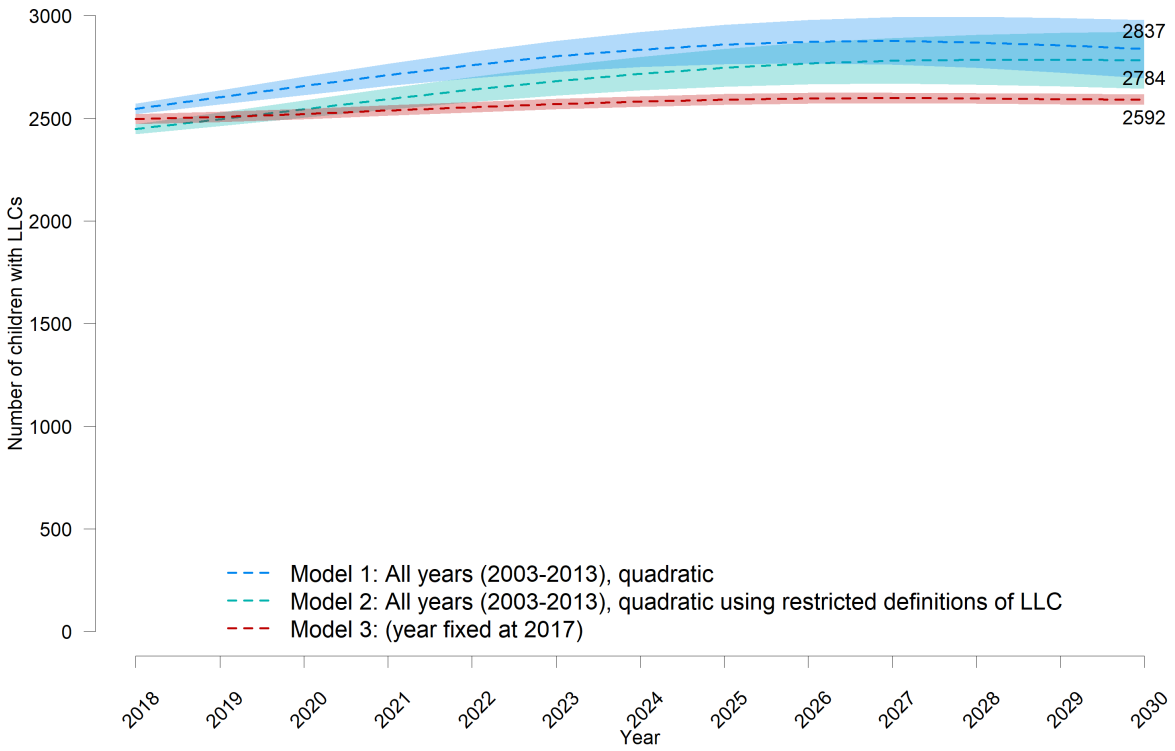


***Figure 28: Predicted number (with 95% confidence intervals in lighter shading) of children with a life-limiting condition (age 0-19) in Northern Ireland***

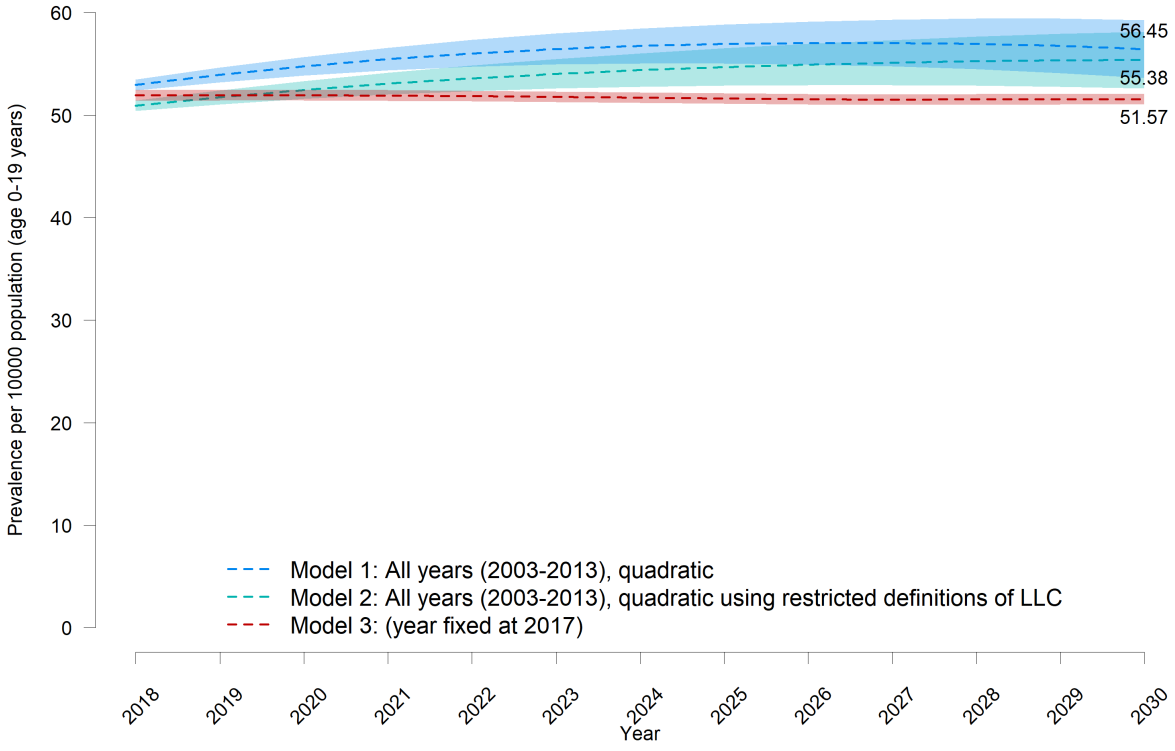


***Figure 29: Predicted prevalence (with 95% confidence intervals in lighter shading) of children with a life-limiting condition (age 0-19) in Northern Ireland***

## Discussion

There have been marked increases in the absolute numbers of children with a LLC in England identified using this methodology. The absolute number of children with a life-limiting condition has risen by 2.6 times in the past 17 years. The rate of increase was greater up to 2010, being twofold, than the rise after 2010 (~1.4 times). This increase is mirrored by a similar rise in prevalence of ~2.5 times (1.8x up to 2010, 1.3x since 2010). Restricting the included diagnoses did not make a large impact on the prevalence (reduced from 66.4 per 10,000 to 63.2 per 10,000) although the effect on the overall prevalence does increase over time.

Data from Scotland, using the same methodology as the current study but including young people up to 21 years of age, has shown a less marked rise in prevalence and no increase from 2014/15-2016/17 (27). This is despite the prevalence in 2003/4 being very similar in Scotland and England (31.5 per 10,000 population vs 32.2 per 10,000 population). Whilst the different demographics of the population, especially ethnic minority populations (13), may partly explain this difference, there may also be differences in healthcare coding practices.

The largest increase in prevalence is seen in the under 1 age group, although it is not clear if this is a change in recording/coding practice or a true increase in diagnoses of LLC in the under one age group. Previous research has shown that the prevalence in children under 1 was initially lower in Scotland in 2003/04 (152.4 per 10,000 population vs 134.2 per 10,000 population) although the prevalence dropped over the subsequent 13 years to 124.1 per 10,000 population (2016/17). However, using a similar methodology the prevalence in England increased to 235.7 per 10,000 population.

The higher prevalence in boys than girls is similar to previous studies (2,3)(28).

The prevalence and increase in prevalence is greatest for congenital disorders, followed by perinatal disorders which partially explains why the increase in prevalence is greatest in the under 1 age group.

The prevalence of LLCs is greater among those of Pakistani, Other Asian and Black ethnic minority groups compared to the White population. Previous literature has confirmed that some of the LLC diagnoses, including some genetic conditions and congenital disorders (29, 30), are more common in those of Pakistani origin.

More children with a LLC lived in areas of higher deprivation which is important when planning services and accessibility of those services. There is some evidence that the proportion of children living in areas of higher deprivation is increasing over time.

Whether there has been a true increase in incidence in LLCs is more difficult to ascertain from these data. Whilst there is evidence of increased recording of LLC diagnoses in the under 1 year age group, it is not possible to differentiate between true increase in the number of children being diagnosed and changes in coding practices. Given the number of electronic medical recording systems being introduced in the NHS the latter cannot be discounted.

There is no evidence that individual children with a LLC are having more hospital admissions and therefore having more opportunity to be recorded within the datasets. Despite an increase in the number of hospital admissions for the population of children with LLCs, the average number of admissions per child remains constant. This is in contrast to the general trend of increasing admissions in children in England (31, 32).

These analyses provide some evidence to support that the increasing prevalence is in part being driven by increased survival. The proportion of children with LLCs who died declined over time and the number who are reaching age 20 years, and may require services from adult healthcare, is increasing.

A hospital admission greater than 28 days may be a poor proxy for complexity; detailed information on technology dependencies and needs of the child would be more helpful (33). The proportion of children with a LLC that had a length of stays >28 days was relatively static at 1.9-2.5% of all admissions or 4.9-5.5% of all admissions with an overnight stay but the absolute number did increase by 1,000 from 2,482 in 2001 to 3,538 in 2017. This increase in absolute number will impact on health services.

Given that the predictions of future prevalence are based on previous trends and predicted changes in population, it is unsurprising that the estimates of future numbers and prevalence shows a continual increase with an estimated prevalence of between 67.0 and 84.2 per 10,000 population.

## Strengths and Limitations

This study used a transparent and repeatable methodology over a long time period which enables assessment of change over time.

Case identification: There is large variation in the severity and prognosis within some of these diagnoses. This makes it challenging to quantify the needs of the child purely by their diagnoses. This is compounded by the grouping of some diagnoses within ICD-10, i.e. each diagnoses does not have its own code.

As a child was only required to have one recording of a LLC or LTC to be included in these analyses, we may have included individuals who had a life-threatening event, i.e. around the time of birth, but who are no longer considered as having an LTC or LLC.

Direct comparison with the previous study from England is not possible as in this study children were included if they had a hospital admission for any cause after a LLC was recorded. The previous study had lower numbers for the same time period due to only including hospital admission recorded with a LLC (7). However the current study does include the whole time period and so trends can still be assessed.

The hospital data used in this study was primarily collected for financial purposes, rather than for research. However, the key variable for this study was the diagnostic code, which is mandatory for financial purposes and therefore collected to a high quality. Some of the other

variables, such as ethnicity, are less well recorded.

If a child did not have a hospital admission for a particular year, they were not included in the data set for that financial year resulting in an underestimation of the numbers of children alive with a LLC for that year.

A projections exercise such as this, involves substantial assumptions about the similarity of future and past trends and also about the future trends in health improvement that may or may not be true. Predicting future prevalence split by age category gave unstable results. Alongside any uncertainties in the numbers diagnosed, the population projections used as denominators are subject to variation from reality, due to variations in demographic rates in the future.

## Conclusions

The prevalence of children with a LLC has increased markedly over this 17 year study period, which can be partially attributed to increased survival and earlier recording of these diagnoses. The prevalence is by far the greatest in children under 1 year of age, as is the number of deaths. This group should be seen as a priority for receiving palliative care as mortality rate is also highest in the under 1 age group. The number of young people surviving paediatric services is increasing and as there are a large number of deaths in those in their 20s, these young people will also require access to appropriate services.

Further research is required to identify the needs and complexity of these children which go beyond their underlying diagnoses. This can only be resolved by more specific coding systems and recording of needs rather than diagnoses alone.

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# Appendix 1-Methods

## Modelling of future prevalence

### England

The HES data from 2004-2016 were used to develop this model as there were more missing data for ethnicity prior to 2004. Additionally, the payment by results system was introduced in England in 2004 under which commissioners pay healthcare providers for each patient seen or treated, taking into account the complexity of the patient's healthcare needs, which may have effected coding practices (14).

The annual probability of an individual having a LLC was estimated using logistic regression. Age categories, sex, ethnicity, GOR, and year were included as predictive variables. The regression equation used was:

$$\log_e\left(\frac{P}{1-P}\right) = C + \beta_1 x_i + \cdots \beta_i x_i + \beta_{y1} year + \beta_{y2} year^2$$

Where *P* is the probability of an individual having a LLC and $x_i$ are binary the variable female and categorical variables age group, ethnic group and GOR.

First, a dataset was generated for each year containing counts of individuals in the national population with and without a LLC for all possible unique combinations of the following demographics: sex, age group, ethnicity and GOR.

The regression was then run to estimate the probability of an individual, for each unique demographic combination, having a LLC in that year. The regression equation was then applied to predict (from Ethpop data (18)) numbers with each unique demographic combination of individuals in years 2018-2030 to predict the probability of a LLC for each individual. The predicted number of individuals with a LLC was then estimated by multiplying the probability of having a LLC by the total estimated number of children with that unique combination of demographics from the Ethpop data. These were then summed to give annual totals of expected individuals with LLC across all demographic combinations.

$$N_{LLC} = \sum_d P_{LLC,d} \times N_d$$

Where $N_{LLC}$ is the annual predicted number of individuals with a LLC; $P_d$ is the probability of an individual in unique demographic combination group (d) having a LLC and $N_d$ is the number of individuals (from Ethpop data) predicted to be in that unique demographic group.

The year terms in the model reflect changes in probability of an individual having a LLC not explained by demographics, i.e. increases in survival and/or incidence rates of LLC over time. Inclusion of a linear year term alone would result in predicted numbers with LLC being forced to be monotonic with year (i.e. always increasing or always decreasing). Hence a quadratic year term was included.

The above approach was used to estimate two models, with different input data:

The first (**Model 1**), used estimates of numbers of individuals with LLC from 2004-2016 and the second, **(Model 2**) used estimates from the same time period but where the restricted definition of a LLC was applied (i.e. excluding oncology cases 5 years after 1st diagnosis and perinatal diagnosis 1 year after birth).

Given the uncertainty of any further improvement in survival or increase in incidence, a further model **(Model 3)** was created, in which all future numbers with a LLC were predicted using a value of 2017 for year, showing predicted numbers if there was no further change in survival and incidence.

The predicted prevalence (per 10,000) of children with LLCs was calculated by dividing the predicted number of children with LLCs by the total population estimate and multiplying the result by 10,000.

Predictions of future prevalence were also made stratified by GOR. This was done using **Model 1.**

Estimations were also made to predict the numbers of children with a LLC in each of the diagnostic groups. For the years 2004-2016, a separate model was developed for each of the eleven diagnostic groups, estimating the probability of an individual who did have a LLC (in any group) having a LLC in that specific diagnostic group (under the restricted definition, i.e. excluding oncology cases 5 years after 1st diagnosis and perinatal diagnosis 1 year after birth). The predictors were again age categories, sex, ethnicity, GOR, and year. Year was only included in its linear form as, due to the smaller numbers per diagnostic group and consequent variation between years, the models became unstable (i.e. output varied greatly with small variations in specification or data used) with the inclusion of a quadratic year term. The predicted probability of having a LLC in a specific diagnostic group was then multiplied by the predicted number (from **Model 2**) having any LLC to give the predicted number of individuals with a LLC in that diagnostic group.

$$N_{Oncology} = \sum_{d} P_{oncology,d} \times P_{LLC,d} \times N_d$$

## Scotland, Wales and Northern Ireland

A similar approach was used with Scottish data previously used for the ChiSP project (8). The annual probability of an individual in Scotland having an LLC was estimated using logistic regression as for the English data, except that only age group, sex and the year terms were included as Ethpop data did not provide regional estimates for Scotland and there were large numbers of individuals missing ethnic group information (22.6% overall). The logistic regression was then applied to Ethpop data for years from 2018 to 2030. As for the English data, three models were applied, one using an unrestricted definition of life-limiting conditions, one using the restricted definition and one using a value of 2017 for all year terms, showing what would happen if there were no further changes in incidence or survival.

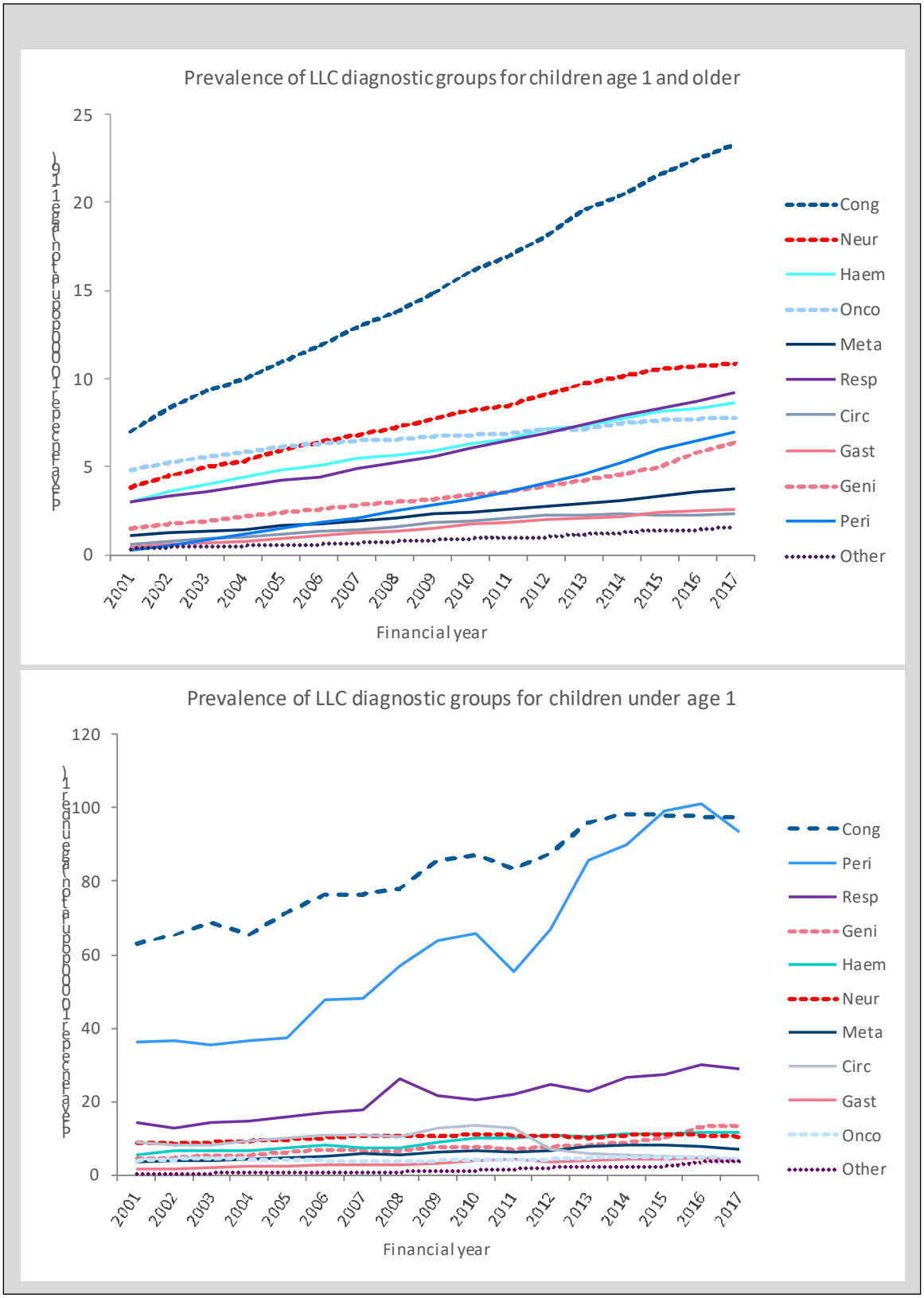
For Wales and Northern Ireland, as there were no individual level data available to build regression models, future numbers of individuals with a LLC were estimated using the Scottish regression models, in preference to the English models as the demographic characteristics of the Welsh and Northern Irish populations were considered closer to those of Scotland than of England. The Scottish regression models were applied to Ethpop data for Wales and Northern Ireland

# Appendix 2-Supplementary Tables and Graphs

***Supplementary Table 1: Number of diagnostic groups per child by financial year***

| Number of Diagnostic groups | 1 | 2 | 3 | 4 | 5 | Total | % with >1 |
|---|---|---|---|---|---|---|---|
| 2001/02 | 27,017 | 4,592 | 478 | 66 | 16 | 32,975 | 15.6% |
| 2002/03 | 29,475 | 5,551 | 680 | 98 | 19 | 36,689 | 17.3% |
| 2003/04 | 31,548 | 6,312 | 895 | 146 | 31 | 39,819 | 18.5% |
| 2004/05[1] | 32,821 | 7,058 | 1,096 | 185 | 43 | 42,114 | 19.9% |
| 2005/06 | 35,441 | 7,953 | 1,334 | 220 | 53 | 45,974 | 20.8% |
| 2006/07 | 37,573 | 8,878 | 1,514 | 290 | 56 | 49,285 | 21.8% |
| 2007/08 | 39,689 | 9,723 | 1,775 | 351 | 96 | 52,633 | 22.7% |
| 2008/09 | 42,380 | 10,586 | 1,986 | 387 | 104 | 56,436 | 23.1% |
| 2009/10 | 44,393 | 11,453 | 2,330 | 492 | 131 | 59,851 | 24.1% |
| 2010/11 | 46,476 | 12,292 | 2,623 | 612 | 180 | 63,254 | 24.8% |
| 2011/12 | 46,614 | 12,805 | 2,996 | 699 | 210 | 64,420 | 25.9% |
| 2012/13 | 49,805 | 13,750 | 3,315 | 800 | 251 | 69,035 | 26.2% |
| 2013/14 | 53,129 | 14,530 | 3,607 | 913 | 322 | 73,608 | 26.3% |
| 2014/15 | 55,384 | 15,212 | 3,972 | 1,054 | 392 | 77,164 | 26.7% |
| 2015/16 | 57,884 | 16,158 | 4,371 | 1,218 | 440 | 81,171 | 27.3% |
| 2016/17 | 59,556 | 16,821 | 4,759 | 1,404 | 509 | 84,267 | 27.9% |
| 2017/18 | 60,748 | 17,469 | 5,043 | 1,550 | 561 | 86,624 | 28.4% |

[1] Introduction of payment by results




***Supplementary figure 1: Comparison of prevalence of life-limiting conditions per 10,000 between children >1 & <1 by diagnostic group***

***Supplementary Table 2***

**Annual numbers of children age 0-19 years with a life limiting condition1 by Government office region for financial years 2000/01 – 2017/18**

| | Financial Year | | | | | | | | | | | | | | | | | |
|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|
| | 2000 | 2001 | 2002 | 2003 | 2004 | 2005 | 2006 | 2007 | 2008 | 2009 | 2010 | 2011 | 2012 | 2013 | 2014 | 2015 | 2016 | 2017 |
| **East Midlands** | 2455 | 2718 | 3020 | 3215 | 3273 | 3599 | 3950 | 4215 | 4421 | 4777 | 4938 | 4971 | 5236 | 5468 | 5719 | 6008 | 6178 | 6655 |
| **East of England** | 2929 | 3313 | 3728 | 4098 | 4251 | 4599 | 4842 | 5360 | 5539 | 6002 | 6462 | 6672 | 7287 | 7637 | 8101 | 8492 | 8884 | 8833 |
| **London** | 4430 | 4921 | 5457 | 5918 | 6373 | 7174 | 7642 | 8336 | 9002 | 9485 | 10113 | 10545 | 11403 | 12421 | 13094 | 13608 | 14083 | 14360 |
| **North East** | 1543 | 1924 | 2092 | 2256 | 2416 | 2642 | 2756 | 2982 | 3127 | 3228 | 3338 | 3334 | 3455 | 3587 | 3745 | 3900 | 3998 | 4054 |
| **North West** | 4554 | 5149 | 5734 | 6174 | 6438 | 6848 | 7526 | 7874 | 8604 | 8747 | 9297 | 9362 | 9908 | 10468 | 11047 | 11230 | 11765 | 12279 |
| **South East** | 4574 | 5157 | 5760 | 6186 | 6542 | 7222 | 7583 | 8160 | 8640 | 9271 | 9791 | 9822 | 10541 | 11284 | 11708 | 12321 | 13014 | 13489 |
| **South West** | 2784 | 3179 | 3505 | 3880 | 4112 | 4505 | 4751 | 4865 | 5465 | 5820 | 6142 | 6179 | 6301 | 6837 | 6989 | 7543 | 7933 | 7954 |
| **West Midlands** | 4436 | 3315 | 3631 | 4134 | 4552 | 4948 | 5372 | 5730 | 6151 | 6748 | 7097 | 7135 | 7845 | 8462 | 8845 | 9529 | 9628 | 9777 |
| **Yorkshire & Humber** | 2830 | 3299 | 3761 | 3958 | 4157 | 4437 | 4863 | 5111 | 5487 | 5773 | 6078 | 6400 | 7060 | 7444 | 7915 | 8541 | 8787 | 9224 |

[1] Numbers are based on all hospital admissions subsequent to a diagnosis for a life-limiting condition

***Supplementary Table 3***

**:** Annual prevalence of children age 0-19 years with a life limiting condition[1] by Government office region for financial years 2001/02 – 2017/18

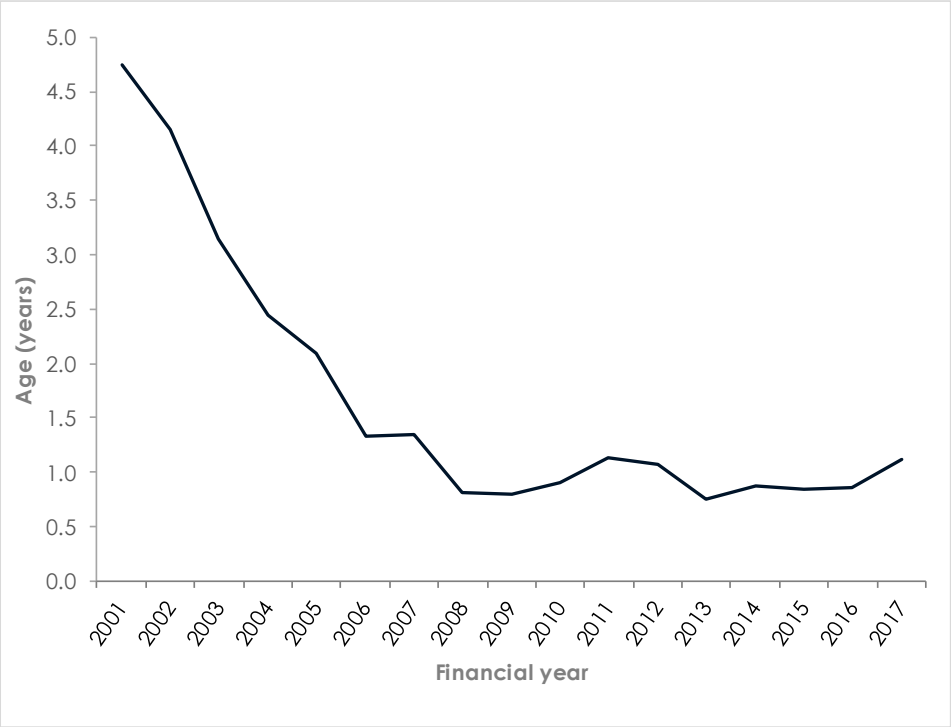
| | Financial Year | | | | | | | | | | | | | | | | |
|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|
| | 2001 | 2002 | 2003 | 2004 | 2005 | 2006 | 2007 | 2008 | 2009 | 2010 | 2011 | 2012 | 2013 | 2014 | 2015 | 2016 | 2017 |
| **East Midlands** | 25.99 | 28.80 | 30.57 | 31.03 | 34.14 | 37.40 | 39.71 | 41.47 | 44.59 | 45.95 | 46.20 | 48.63 | 50.71 | 53.00 | 55.46 | 56.80 | 60.93 |
| **East of England** | 24.83 | 27.83 | 30.40 | 31.39 | 33.81 | 35.49 | 39.07 | 40.17 | 43.33 | 46.42 | 47.75 | 51.95 | 54.30 | 57.50 | 59.88 | 62.26 | 61.50 |
| **London** | 27.55 | 30.25 | 32.48 | 34.77 | 38.83 | 40.98 | 44.06 | 46.88 | 48.54 | 50.99 | 52.50 | 55.86 | 59.96 | 62.28 | 63.80 | 65.11 | 65.45 |
| **North East** | 30.41 | 33.29 | 36.22 | 39.08 | 43.12 | 45.29 | 49.19 | 51.77 | 53.65 | 55.57 | 55.73 | 58.21 | 60.72 | 63.64 | 66.32 | 68.07 | 69.09 |
| **North West** | 29.57 | 33.05 | 35.67 | 37.33 | 39.92 | 44.03 | 46.16 | 50.56 | 51.47 | 54.85 | 55.39 | 58.75 | 62.11 | 65.58 | 66.55 | 69.62 | 72.56 |
| **South East** | 26.05 | 29.09 | 31.11 | 32.76 | 36.09 | 37.74 | 40.26 | 42.39 | 45.21 | 47.39 | 47.24 | 50.56 | 53.98 | 55.88 | 58.54 | 61.56 | 63.53 |
| **South West** | 27.12 | 29.85 | 32.93 | 34.85 | 38.13 | 40.24 | 41.02 | 46.04 | 49.01 | 51.58 | 51.79 | 52.80 | 57.27 | 58.55 | 63.02 | 66.14 | 66.18 |
| **West Midlands** | 24.30 | 26.63 | 30.28 | 33.33 | 36.22 | 39.26 | 41.73 | 44.58 | 48.69 | 51.08 | 51.29 | 56.23 | 60.39 | 62.90 | 67.52 | 67.96 | 68.75 |
| **Yorkshire & Humber** | 25.96 | 29.68 | 31.24 | 32.83 | 35.07 | 38.54 | 40.46 | 43.32 | 45.39 | 47.66 | 50.12 | 55.27 | 58.19 | 61.76 | 66.33 | 67.99 | 71.06 |

[1] Numbers are based on all hospital admissions subsequent to a diagnosis for a life-limiting condition

Supplementary Table 4: Sensitivity analysis of annual prevalence (per 10,000 population) of children (0-19 years) with life-limiting conditions in England (with restricted definitions of LLCs)

| | Prevalence per 10,000 population | | | | | | | | | | | |
|---|---|---|---|---|---|---|---|---|---|---|---|---|
| Year | Without restrictions | 95% CI | Perinatal < 1 yr old | 95% CI | Oncology < 5yrs | 95% CI | Non CNS-oncology <5yrs | 95% CI | No early stage renal | 95% CI | All restrictions combined | 95% CI |
| 2001/02 | 26.7 | 26.5-27.0 | 26.6 | 26.3-26.9 | 24.8 | 24.5-25.1 | 24.8 | 24.5-25.1 | 26.7 | 26.5-27.0 | 24.7 | 24.4-25.0 |
| 2002/03 | 29.7 | 29.4-30.0 | 29.4 | 29.1-29.7 | 26.7 | 26.4-27.0 | 26.7 | 26.4-27.0 | 29.7 | 29.4-30.0 | 26.6 | 26.3-26.9 |
| 2003/04 | 32.2 | 31.9-32.5 | 31.7 | 31.4-32.0 | 29.6 | 29.3-30.0 | 29.6 | 29.3-30.0 | 32.2 | 31.9-32.5 | 29.3 | 29.0-29.6 |
| 2004/05 | 34.0 | 33.7-34.3 | 33.3 | 33.0-33.6 | 32.1 | 31.8-32.5 | 32.1 | 31.8-32.5 | 34.0 | 33.7-34.3 | 31.6 | 31.3-32.0 |
| 2005/06 | 37.1 | 36.7-37.4 | 36.2 | 35.9-36.6 | 34.0 | 33.6-34.3 | 34.0 | 33.6-34.3 | 37.1 | 36.7-37.4 | 33.3 | 33.0-33.6 |
| 2006/07 | 39.7 | 39.3-40.0 | 38.7 | 38.3-39.0 | 36.8 | 36.5-37.2 | 36.9 | 36.5-37.2 | 39.7 | 39.3-40.0 | 36.0 | 35.6-36.3 |
| 2007/08 | 42.2 | 41.8-42.5 | 41.1 | 40.7-41.4 | 39.1 | 38.7-39.5 | 39.2 | 38.9-39.6 | 42.2 | 41.8-42.5 | 38.1 | 37.7-38.4 |
| 2008/09 | 45.0 | 44.6-45.4 | 43.7 | 43.3-44.1 | 41.5 | 41.1-41.9 | 41.7 | 41.3-42.1 | 45.0 | 44.6-45.4 | 40.4 | 40.1-40.8 |
| 2009/10 | 47.5 | 47.1-47.9 | 46.0 | 45.7-46.4 | 44.3 | 43.9-44.6 | 44.5 | 44.1-44.8 | 47.5 | 47.1-47.8 | 43.0 | 42.6-43.4 |
| 2010/11 | 49.9 | 49.5-50.3 | 48.4 | 48.0-48.7 | 46.7 | 46.3-47.1 | 46.9 | 46.5-47.3 | 49.9 | 49.5-50.3 | 45.3 | 44.9-45.7 |
| 2011/12 | 50.7 | 50.3-51.1 | 48.9 | 48.6-49.3 | 49.2 | 48.8-49.6 | 49.5 | 49.1-49.8 | 50.7 | 50.3-51.1 | 47.7 | 47.3-48.0 |
| 2012/13 | 54.1 | 53.7-54.5 | 52.2 | 51.8-52.6 | 50.0 | 49.6-50.4 | 50.2 | 49.8-50.6 | 54.1 | 53.7-54.5 | 48.2 | 47.8-48.6 |
| 2013/14 | 57.5 | 57.1-57.9 | 55.4 | 55.0-55.8 | 53.4 | 53.0-53.8 | 53.6 | 53.2-54.0 | 57.5 | 57.1-57.9 | 51.4 | 51.0-51.8 |
| 2014/15 | 60.1 | 59.7-60.5 | 57.6 | 57.2-58.1 | 56.8 | 56.4-57.2 | 57.0 | 56.6-57.5 | 60.0 | 59.6-60.5 | 54.6 | 54.2-55.1 |
| 2015/16 | 62.9 | 62.4-63.3 | 60.1 | 59.6-60.5 | 59.3 | 58.8-59.7 | 59.5 | 59.0-59.9 | 62.8 | 62.4-63.3 | 56.8 | 56.4-57.2 |
| 2016/17 | 64.9 | 64.5-65.4 | 62.0 | 61.6-62.4 | 62.1 | 61.6-62.5 | 62.3 | 61.8-62.7 | 64.9 | 64.4-65.3 | 59.2 | 58.8-59.6 |
| 2017/18 | 66.4 | 66.0-66.8 | 63.2 | 62.8-63.6 | 64.1 | 63.6-64.5 | 64.3 | 63.8-64.7 | 66.3 | 65.9-66.7 | 61.1 | 60.7-61.5 |

Abbreviations: 95% CI- 95% Confidence intervals-




***Supplementary figure 2: Median age of first recorded life-limiting diagnosis for children (age 0-19) for 2000/01-2017/18***

Supplementary Table 5: Numbers who died from each birth cohort

| year | Died | | Total | % died |
|---|---|---|---|---|
| | yes | no | | |
| **2001** | 1,238 | 6,016 | 7,254 | 17% |
| **2002** | 1,165 | 6,169 | 7,334 | 16% |
| **2003** | 1,226 | 6,504 | 7,730 | 16% |
| **2004** | 1,296 | 6,725 | 8,021 | 16% |
| **2005** | 1,288 | 7,322 | 8,610 | 15% |
| **2006** | 1,539 | 8,356 | 9,895 | 16% |
| **2007** | 1,485 | 8,712 | 10,197 | 15% |
| **2008** | 1,563 | 10,290 | 11,853 | 13% |
| **2009** | 1,646 | 10,760 | 12,406 | 13% |
| **2010** | 1,566 | 11,180 | 12,746 | 12% |
| **2011** | 1,248 | 10,614 | 11,862 | 11% |
| **2012** | 1,335 | 11,636 | 12,971 | 10% |
| **2013** | 1,444 | 12,965 | 14,409 | 10% |
| **2014** | 1,439 | 13,287 | 14,726 | 10% |
| **2015** | 1,393 | 14,098 | 15,491 | 9% |
| **2016** | 1,325 | 14,589 | 15,914 | 8% |
| **2017** | 1,091 | 14,397 | 15,488 | 7% |
| Total | 23,287 | 173,620 | 196,907 | |

Supplementary Table 6: Numbers who died from each first year of diagnosis cohort

| year | Died | | Total | % died |
|---|---|---|---|---|
| | yes | no | | |
| **2001** | 2,913 | 15,083 | 17,996 | 16.2% |
| **2002** | 2,439 | 14,330 | 16,769 | 14.5% |
| **2003** | 2,264 | 13,772 | 16,036 | 14.1% |
| **2004** | 2,122 | 13,363 | 15,485 | 13.7% |
| **2005** | 2,052 | 14,201 | 16,253 | 12.6% |
| **2006** | 2,161 | 14,856 | 17,017 | 12.7% |
| **2007** | 2,069 | 15,728 | 17,797 | 11.6% |
| **2008** | 2,088 | 16,780 | 18,868 | 11.1% |
| **2009** | 2,131 | 17,409 | 19,540 | 10.9% |
| **2010** | 2,026 | 18,318 | 20,344 | 10.0% |
| **2011** | 1,654 | 18,235 | 19,889 | 8.3% |
| **2012** | 1,640 | 19,894 | 21,534 | 7.6% |
| **2013** | 1,750 | 20,706 | 22,456 | 7.8% |
| **2014** | 1,738 | 21,537 | 23,275 | 7.5% |
| **2015** | 1,645 | 22,886 | 24,531 | 6.7% |
| **2016** | 1,502 | 23,410 | 24,912 | 6.0% |
| **2017** | 1,224 | 23,914 | 25,138 | 4.9% |
| Total | 37,424 | 322,210 | 359,634 | |

Supplementary Table 7: Number of hospital admissions by length of stay for children (aged 0-19) with life-limiting conditions between 2001/02-2017/18

| Financial year | daycase | 1-13 nights | 14-27 nights | ≥ 28 nights | >=56 nights |
|---|---|---|---|---|---|
| 2001 | 77,465 | 65,303 | 6,005 | 2,482 | 1,405 |
| 2002 | 87,249 | 70,554 | 6,077 | 2,470 | 1,513 |
| 2003 | 91,446 | 73,731 | 6,423 | 2,648 | 1,719 |
| 2004 | 97,320 | 75,251 | 6,353 | 2,785 | 1,828 |
| 2005 | 107,679 | 78,485 | 6,827 | 2,854 | 1,805 |
| 2006 | 113,515 | 79,925 | 6,898 | 2,938 | 1,984 |
| 2007 | 122,879 | 81,197 | 7,053 | 3,042 | 1,986 |
| 2008 | 125,010 | 84,170 | 7,266 | 3,001 | 2,152 |
| 2009 | 139,018 | 89,460 | 7,666 | 3,311 | 2,365 |
| 2010 | 144,454 | 93,833 | 7,765 | 3,394 | 2,286 |
| 2011 | 150,395 | 94,934 | 7,572 | 3,310 | 2,351 |
| 2012 | 158,763 | 97,168 | 7,845 | 3,500 | 2,488 |
| 2013 | 165,519 | 98,729 | 8,034 | 3,650 | 2,517 |
| 2014 | 168,067 | 102,547 | 8,065 | 3,610 | 2,698 |
| 2015 | 179,359 | 106,318 | 8,186 | 3,689 | 2,757 |
| 2016 | 183,133 | 107,888 | 8,092 | 3,878 | 2,844 |
| 2017 | 189,151 | 109,417 | 7,693 | 3,538 | 2,476 |

Supplementary Table 8: Hierarchical Logistic regression model (dependent variable LOS ≥ 28 days) excluding birth admission

| | Odds Ratio | SE | Z | P>[z] | 95% Confidence Interval | |
|---|---|---|---|---|---|---|
| Year | 0.97 | 0.00 | -20.74 | 0.00 | 0.97 | 0.98 |
| **Sex** | | | | | | |
| Male | 1.00 (reference) | | | | | |
| Female | 1.06 | 0.01 | 5.26 | 0.00 | 1.04 | 1.08 |
| **Age group** | | | | | | |
| age < 1 | 7.37 | 0.01 | 142.45 | <0.01 | 7.17 | 7.58 |
| age 1-5 | 1.00 (reference) | | | | | |
| age 6-10 | 0.71 | 0.02 | -17.88 | <0.01 | 0.69 | 0.74 |
| age 11-15 | 0.94 | 0.02 | -3.36 | <0.01 | 0.91 | 0.97 |
| age 16-19 | 1.23 | 0.02 | 11.34 | <0.01 | 1.19 | 1.28 |
| **Ethnic group** | | | | | | |
| White | 1.00 (reference) | | | | | |
| Missing | 0.77 | 0.04 | -6.88 | <0.01 | 0.71 | 0.83 |
| Indian | 1.10 | 0.04 | 2.66 | 0.01 | 1.02 | 1.18 |
| Pakistani | 1.25 | 0.02 | 9.63 | <0.01 | 1.19 | 1.31 |
| Bangladeshi | 1.38 | 0.04 | 7.33 | <0.01 | 1.27 | 1.50 |
| Black | 1.44 | 0.02 | 15.24 | <0.01 | 1.38 | 1.51 |
| Chinese | 1.43 | 0.09 | 4.08 | <0.01 | 1.20 | 1.70 |
| Mixed | 1.15 | 0.03 | 4.41 | <0.01 | 1.08 | 1.22 |
| Other Asian | 1.48 | 0.02 | 15.62 | <0.01 | 1.41 | 1.55 |
| **GOR** | | | | | | |
| London | 1.00 (reference) | | | | | |
| East Midlands | 1.03 | 0.02 | 1.31 | 0.19 | 0.98 | 1.08 |
| East of England | 0.92 | 0.02 | -3.45 | 0.00 | 0.88 | 0.97 |
| North East | 0.87 | 0.03 | -4.8 | 0.00 | 0.82 | 0.92 |
| North West | 1.02 | 0.02 | 1.08 | 0.28 | 0.98 | 1.06 |
| South East | 0.98 | 0.02 | -1.07 | 0.28 | 0.94 | 1.02 |
| South West | 0.93 | 0.02 | -3.12 | 0.00 | 0.89 | 0.97 |
| West Midlands | 0.94 | 0.02 | -2.92 | 0.00 | 0.90 | 0.98 |
| Yorkshire & Humber | 0.93 | 0.02 | -3.17 | 0.00 | 0.89 | 0.97 |
| **Deprivation** | | | | | | |
| imd1 (least deprived) | 0.81 | 0.02 | -10.78 | <0.01 | 0.78 | 0.85 |
| imd2 | 0.86 | 0.02 | -8.27 | <0.01 | 0.83 | 0.89 |
| imd3 | 0.88 | 0.02 | -7.89 | <0.01 | 0.85 | 0.91 |
| imd4 | 0.93 | 0.02 | -4.74 | <0.01 | 0.90 | 0.96 |
| imd 5(most deprived) | 1.00 (reference) | | | | | |
| **Diagnoses** | | | | | | |
| Congenital | 1.00 (reference) | | | | | |
| Circulatory | 1.90 | 0.03 | 19.17 | <0.01 | 1.78 | 2.03 |
| Gastrointestinal | 3.13 | 0.04 | 25.55 | <0.01 | 2.87 | 3.42 |
| Genitourinary | 2.27 | 0.03 | 31.47 | <0.01 | 2.16 | 2.39 |
| Haematology | 1.67 | 0.03 | 19.72 | <0.01 | 1.59 | 1.76 |

| | | | | | | |
|---|---|---|---|---|---|---|
| Metabolic | 1.15 | 0.03 | 4.31 | <0.01 | 1.08 | 1.22 |
| Neurology | 1.79 | 0.02 | 30.7 | <0.01 | 1.73 | 1.86 |
| Oncology | 3.33 | 0.02 | 67.93 | <0.01 | 3.21 | 3.44 |
| Other | 2.09 | 0.04 | 16.83 | <0.01 | 1.92 | 2.27 |
| Perinatal | 0.79 | 0.02 | -11.01 | <0.01 | 0.76 | 0.83 |
| Respiratory | 1.59 | 0.02 | 23.71 | <0.01 | 1.53 | 1.65 |
| **Emergency Admission** | | | | | | |
| Non-emergency emission | 1.00 (reference) | | | | | |
| Emergency | 0.98 | 0.01 | -2.35 | 0.02 | 0.96 | 1.00 |

Supplementary Table 9: Numbers of children who survive to age 19

| Financial year | Number of children with LLCs aged 19 | Number who died | | Numbers who survived | |
|---|---|---|---|---|---|
| | | N | % | N | % |
| **2001** | 990 | 69 | 7.0% | 921 | 93.0% |
| **2002** | 1,044 | 47 | 4.5% | 997 | 95.5% |
| **2003** | 1,205 | 66 | 5.5% | 1139 | 94.5% |
| **2004** | 1,311 | 54 | 4.1% | 1257 | 95.9% |
| **2005** | 1,403 | 65 | 4.6% | 1338 | 95.4% |
| **2006** | 1,593 | 72 | 4.5% | 1521 | 95.5% |
| **2007** | 1,733 | 63 | 3.6% | 1670 | 96.4% |
| **2008** | 1,889 | 57 | 3.0% | 1832 | 97.0% |
| **2009** | 1,986 | 71 | 3.6% | 1915 | 96.4% |
| **2010** | 2,162 | 68 | 3.1% | 2094 | 96.9% |
| **2011** | 2,172 | 53 | 2.4% | 2119 | 97.6% |
| **2012** | 2,295 | 52 | 2.3% | 2243 | 97.7% |
| **2013** | 2,354 | 51 | 2.2% | 2303 | 97.8% |
| **2014** | 2,534 | 58 | 2.3% | 2476 | 97.7% |
| **2015** | 2,669 | 68 | 2.5% | 2601 | 97.5% |
| **2016** | 2,845 | 55 | 1.9% | 2790 | 98.1% |
| **2017** | 3,131 | 56 | 1.8% | 3075 | 98.2% |