Children with a Life-limiting or Life-threatening condition in Wales: Trends in prevalence and complexity

Final Report

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Authors

Lorna Fraser, Professor of Epidemiology, University of York

Andre Bedendo, Research Fellow, University of York

Stuart Jarvis, Research Fellow, University of York

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Executive summary

- Children and Young people with life-limiting and life-threatening conditions may benefit from input from Paediatric Palliative Care services.
- Effective healthcare planning, service development and delivery relies upon an accurate understanding of the population who would benefit from that care. Although recent studies in England and Scotland have shown an increasing prevalence of children and young people with life-limiting conditions, data in Wales has not been updated since 2009.
- The aim of this report was to summarise the key findings on the population of children and young people in Wales with a life-limiting or life-threatening condition. This included identifying the numbers of children, prevalence by age, deprivation and health board and complexity of needs of this population of children and young people.
- An analysis of routinely collected linked primary and secondary healthcare data was undertaken to provide this information.
- The results of this report are summarised in the following Key Findings:

KEY FINDING 1 - The population of children and young people with life-limiting conditions increased by almost a quarter in the decade from 2009-2019

KEY FINDING 2 - There are geographical disparities in prevalence of life-limiting conditions between health boards and more children and young people living in areas of higher deprivation than expected

KEY FINDING 3 - Congenital conditions have the highest prevalence, but there are marked increases in prevalence in most diagnostic groups

KEY FINDING 4 - Children and young people with life-limiting conditions are disproportionately high users of healthcare

KEY FINDING 5 - There is a range of medical complexity across conditions and increasing complexity with increasing age

KEY FINDING 6 - Approximately a third of children and young people with life-limiting conditions in Wales experience clinical instability each year

KEY FINDING 7 - Medical complexity and stage of condition are related, but complexity provides additional information on needs

KEY FINDING 8 - More than half of children who die in Wales have an underlying life-limiting condition

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Background and context

Effective healthcare planning, service development and delivery relies upon accurate understanding of the population who would benefit from that care. Children and young people with life-limiting or life-threatening conditions (hereafter life-limiting conditions, see box 1) may benefit from input from paediatric palliative care services.(1) Data on this population of children and young people has not been updated since 2009 in Wales.(2) More recent studies from England (3) and Scotland(4) have shown increasing numbers and prevalence of life-limiting conditions in children and young people. These studies have also shown higher prevalence of these conditions in areas of higher deprivation and in ethnic minority populations.

One challenge of the data sources used to date has been the lack of information available on the severity or complexity of the child's life-limiting condition. Wales is in the unique position of having full population linked to primary and secondary care healthcare data (5) which could enable the analyses to go beyond the diagnoses into an assessment of complexity.

Box 1 -Definitions

Life Limiting conditions : are those for which there is no reasonable hope of cure and from which children or young people will die.(6)

Life-threatening conditions: are those for which curative treatment may be feasible but can fail, such as cancer.(6)

This Study

The present study was commissioned by the NHS Wales National Programme Board for Palliative and End of Life Care to provide evidence for future planning of healthcare and support services in Wales for children and young people with life-limiting conditions.

Aim: To update the prevalence of children with a life-limiting condition in Wales and assess the complexity of this population to aid service planning and delivery.

Objectives:

1. To assess trends in the numbers of and prevalence of children with a life-limiting condition in Wales from 2009-2019.
2. To assess what proportion of these children are stable/unstable/deteriorating or dying?
3. To assess the complexity of children with a life-limiting condition and the trends over time in this complexity
4. To describe the trends in the healthcare usage of children with a life-limiting condition

The study utilised routinely collected linked healthcare records in Wales, held securely within the SAIL (Secure Anonymised Information Linkage) Databank[1] at the University of Swansea. SAIL provides high quality) routinely collected healthcare data and many of the data items are recorded for payment purposes, so providers are incentivised to ensure accuracy. The datasets are probabilistically linked using an algorithm shown to have very high matching accuracy.(7-9) This dataset was enhanced by the addition of paediatric

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[1] https://saildatabank.com

Paediatric Palliative Care in Wales

In the trajectory of every new clinical service, there comes a point when it is no longer enough for those responsible for developing and funding it to be motivated by moral conviction, and we need to turn instead to data.

It would be hard to say exactly when that moment arrives. Almost by definition, a novel specialty is novel precisely because no-one has previously thought to look for the evidence that it is necessary. Any new way for healthcare to serve patients starts, not because there is data to suggest it is needed, but because someone has observed that we are not treating patients as well as we should, and on that basis has decided that things should be done differently. The need for a novel service is a moral assertion, based on compassion and common humanity. Evidence, if you like, of a sort; but evidence based on reasoning, not on data.

Several things are needed for a service to get beyond that embryonic stage. First of all, a funder must be found who is willing to support initial development on the basis of conviction alone, until the service finds itself in a position to measure its own increasing activity as its reach extends. Since the Sugar Report in 2009, paediatric palliative care in Wales has enjoyed exactly that kind of support from the Welsh Government and we now use a bespoke suite of metrics and outcome measures to collect data prospectively.

But such ‘in house’ data can only tell us about the children our service is already reaching. As the numbers augment year on year, it becomes impossible to avoid the inference that there are other children whom we are not reaching, and might not even be aware of. It is an inevitable blind spot because, of course, we cannot know how much we don’t know. And that’s where paediatric palliative care in Wales has really fallen on its feet. Thanks again to support from the Welsh Government we have been able to engage the services of Professor Lorna Fraser and her team from the University of York. In the report that follows, Professor Fraser’s research has been able to tell us about other children with life-limiting conditions. The ones even Wales’ well-developed palliative care service doesn’t see. Children whose families do not need specialist palliative medicine, but do need the wider palliative and short break care that children’s hospices can offer. Children who do need specialist palliative care, but cannot access it because of the nature of their diagnosis or where they live. Complex children on dozens of different medications, whose exhausted parents manage medical devices in the home for years or decades. Thanks to this report, we no longer have to guess how many children in Wales might need to access palliative care. We now know that the number is well over four thousand (that’s rather more children than have diabetes), and that the number is rising steadily year on year. We know that children with life-limiting conditions live in every area in Wales; not just those within reach of acute paediatric departments, or with good community children’s nursing support. We know that they are among the most medically complex children in Wales. And we know that, as so often, the burden is falling most heavily on those least able to bear it financially.

That data powerfully informs how we deliver children’s palliative care services, not only in hospital and at home but also in the Children’s Hospices in Wales who can provide the spaces, places and resources that allow children and young people to thrive. The data allows children’s hospices both to refine and to target their services, focusing on areas of Wales where there is most need for their services, and enables them to engage politically by providing a base for discussions with decision- and lawmakers in order to facilitate the best possible service for children and young people with life limiting and life limiting conditions in Wales.

Does all this data mean that paediatric palliative care in Wales has come of age? That we are ready to set aside moral assertions about what is needed and henceforth develop palliative care services for children wholly in response to data? Well, no, not quite yet. For the time being, the argument for development of paediatric palliative care services is still, at least partly, a moral one. We should still be appalled by a degree of inequity in Wales that means a child dying at home from a metabolic disease can only access specialist palliative care nursing between 9 and 5 during the week, while an adult, or a child dying from cancer, can have 24 hour access. We should be outraged that in Betsi Cadwaladr and Aneurin Bevan there are palliative care nurses for adults, but none for children, and that in Powys there is no children’s palliative care provision whatsoever. These are matters of right and wrong; of fair and unfair; of justice and injustice. We do not need data to know they must be urgently redressed.

But a last push to get us to the evidence-base starting line need not take long. Once it is done, the data in this report mean that paediatric palliative care in Wales will at last be able to follow in the footsteps of its sister specialty in adults, and finally move forward in a direction that is shaped, not only by compassion and the demands of justice, but also and increasingly by objective evidence. We may not have reached the starting-blocks quite yet but, thanks to the data in this report, when we do so we will already be running.

Our thanks go to Lorna Fraser and Stuart Jarvis from the University of York for undertaking the huge amount of work needed to complete this project. Thanks must also go to the National Palliative and End of Life Care Programme Board for funding the project, and the steering group for navigating the work through inception to publication.

***Richard Hain** Consultant and Clinical Lead All-Wales Managed Clinical Network in Paediatric Palliative Medicine*

***Andy Goldsmith** CEO Hope House and Tŷ Gobaith*

***Maria Timon Samra** CEO Tŷ Hafan*

across the UK. Data were available up to 2020, but are presented herein to 2019 due to the effects of the COVID-19 pandemic on healthcare use and delivery from early 2020.

With these rich data, we were able to estimate how the size of the population has changed over the decade to 2019 and to look at healthcare use, deaths, medical complexity and stage of condition, to better understand the population, its healthcare use and impact on services in Wales. The population of interest was children aged under 18 years, but the report also includes information on young people under 26 years with conditions diagnosed during childhood (before 18 years) as this population is important in terms of transition to adult services. Hereinafter, this report refers to children (under 18 years - i.e. from birth to 17 years 364 days) and children and young people (under 26 years - i.e. from birth to 25 years 263 days).

The full methods are detailed in the appendix. This study has approval from the Research Ethics Committee and the Confidentiality Advisory Group of the Health Research Authority (refs: 21/LO/0567, 21/CAG/0114).

These results are presented under eight key findings which may be used by the funder, services and policy makers to plan and target services.

Key Finding 1: The population of children with life-limiting conditions increased by almost a quarter in the decade from 2009-2019

Overview of Methods

Datasets

The key datasets for estimating the number of children and young people with a life-limiting conditions in Wales were inpatient records and General Practice records (Figure 1). Inpatient records (from the Patient Episode Dataset for Wales) included information on admission and discharge dates, diagnoses, treatments and area of residence for children and young people admitted to hospitals in Wales. General Practice records (from the Welsh Longitudinal General Practice Dataset) included details of dates of appointments, diagnoses and prescriptions. These were used to identify the presence of life-limiting conditions as set out below and to identify those still present in Wales and requiring inpatient care.

Of note, we requested access to hospital admission data from England recognising that some children who lived on the border with England or in the North of Wales may receive care in the NHS in England. Unfortunately, NHS Digital (now NHS England) were unable to provide these data.

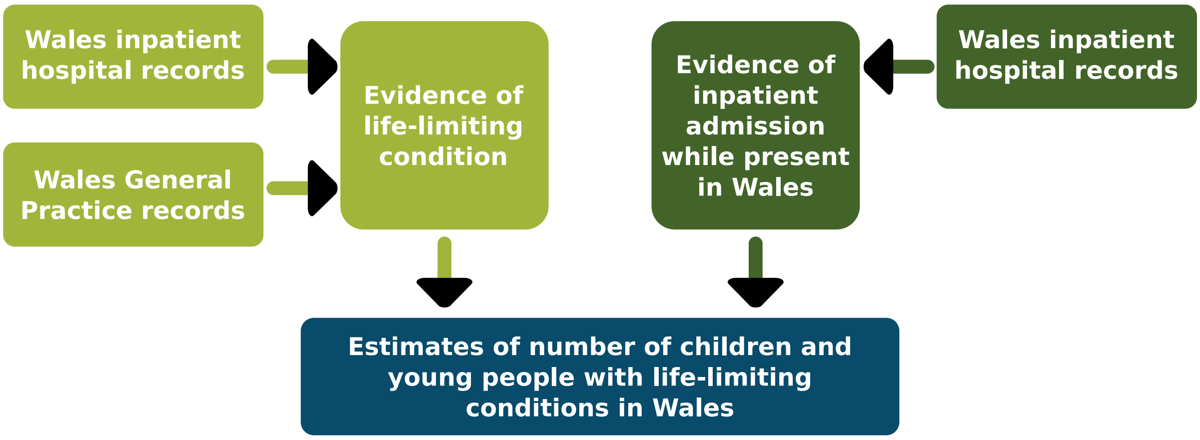

Figure 1: Overview of datasets used to ascertain presence of a life-limiting condition and continued presence in Wales with a need for inpatient care.

Cohort Identification

Children and young people were included in this cohort if they had a record of any of the Life-limiting or life-threatening conditions recorded in their Primary Care record or Hospital admission data whilst aged under 18 years[1].

Children and young people were included in the prevalence estimates in each year if they had a life-limiting condition diagnosis[2] in that year or any previous year *and* had a hospital inpatient admission (of any length, including day cases) in that year.

Numbers of children (under 18 years) and children and young people (under 26 years) with a life-limiting condition and the *prevalence* of life-limiting conditions within the population of these ages in Wales were calculated.

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[2] See appendix for a full list of diagnoses. Diagnoses for perinatal conditions were only included for children aged under 1 year (see appendix for further details).

Prevalence is a measure of the number of individuals with a given condition in a population at a given time. Here it is expressed as the number of individuals in each 10000 children and young people in Wales each year that have a life-limiting condition:

$$prevalence = \frac{number\ of\ individuals\ with\ a\ LLC}{population\ at\ risk}\ x10000$$

Results

The number of children (under 18 years) with a life-limiting condition increased by 22.8% from 2977 in 2009 to 3655 in 2019 (Figure 2, Table 1). For children and young people (under 26 years) diagnosed in childhood, there was a 29.8% increase from 3304 in 2009 to 4289 in 2019.

Prevalence of life-limiting conditions increased from 47 (in 2009) to 58 per 10000 (in 2019) in children (under 18 years) and from 34 to 45 per 10000 in children and young people (under 26 years). Prevalence was highest in children under 1 year, but the largest proportional increases were seen in older age groups, particularly those aged 12-17 years and 18-25 years, likely indicative of increasing survival times (Figure 2, There were more males than females with life-limiting conditions in all years (Table 1).

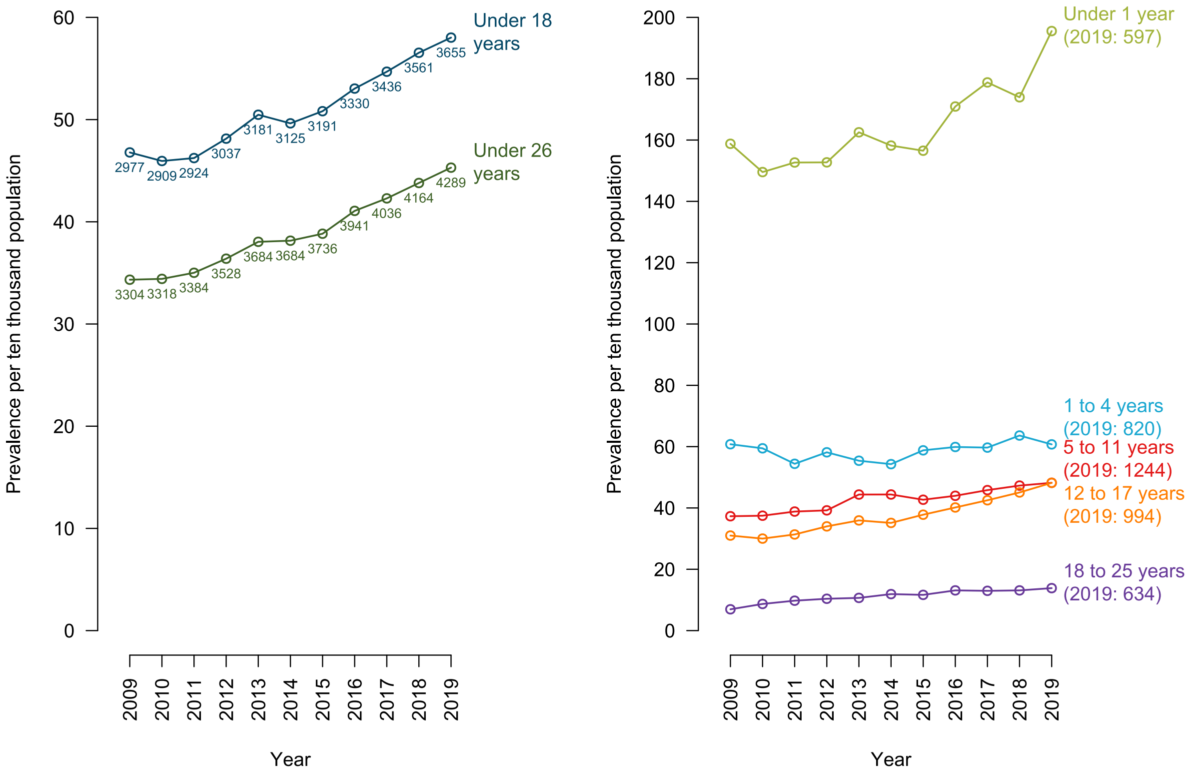


Figure 2: Prevalence of life-limiting conditions recorded in childhood among children and young people in Wales. Left: overall prevalence for children (under 18 years) and children and young people (under 26 years). Figures adjacent to points are numbers of young people. Right: prevalence by age group with numbers for 2019.

Table 1: Numbers of children and young people with life-limiting conditions, by year. Unless otherwise stated, figures are for children and young people aged under 26 years.

	Year										
	2009	**2010**	**2011**	**2012**	**2013**	**2014**	**2015**	**2016**	**2017**	**2018**	**2019**
Age group (years)											
0-17	2977	2909	2924	3037	3181	3125	3191	3330	3436	3561	3655
0-25	3304	3318	3384	3528	3684	3684	3736	3941	4036	4164	4289
Under 1	556	530	555	543	559	532	525	570	581	556	597
1-4	836	833	773	839	800	782	837	838	825	870	820
5-11	873	868	897	913	1050	1071	1052	1108	1173	1219	1244
12-17	712	678	699	742	772	740	777	814	857	916	994
16-17	225	222	235	245	259	255	273	269	259	268	300
18-25	327	409	460	491	503	559	545	611	600	603	634
Gender											
Males	1827	1836	1866	1949	2041	2021	2029	2181	2197	2280	2311
Females	1477	1482	1518	1579	1643	1663	1707	1760	1838	1884	1978
Deprivation category											
1 (most deprived)	818	797	818	801	872	876	869	973	990	994	1084
2	729	743	752	802	845	812	771	866	869	905	904
3	596	573	599	627	660	639	656	692	716	755	760
4	579	631	638	652	620	652	692	700	729	720	757
5 (least deprived)	582	574	577	646	687	705	748	710	732	790	784
Local Health Board											
Betsi Cadwaladr UHB	659	676	721	722	717	750	739	776	805	845	885
Hywel Dda UHB	322	322	331	374	396	407	404	422	430	443	439
Powys THB	88	101	115	115	114	114	122	150	138	127	140
Swansea Bay UHB	390	388	388	388	408	413	428	445	455	476	477
Cwm Taf Morgannwg UHB	529	512	572	609	632	610	601	630	687	711	721
Aneurin Bevan UHB	761	754	709	715	767	781	779	852	837	859	916
Cardiff and Vale UHB	555	565	548	605	650	609	663	666	684	703	711

Key Finding 2: There are geographical disparities in prevalence of life-limiting conditions between health boards and more children and young people living in areas of higher deprivation than expected

Overview of methods

Children and young people with life-limiting conditions were assigned demographic data in each year based on their first record in the available datasets.

Each individual was assigned to a Local Health Board and a deprivation category based on postcode of residence. The deprivation categories used were population-weighted i.e. 20% of all 0-25 year olds in Wales lived in each of the five groups so if life-limiting conditions were equally distributed in the population 20% of those children should also live in each of the five deprivation groups.

Data were of insufficient quality and completeness to assign ethnic group to individuals. Therefore, a population-level assessment was made between the prevalence of life-limiting conditions and the proportion of non-White ethnic groups in each Local Health Board.

Results

At the Local Health Board Level, there was higher prevalence of life-limiting conditions in Cwm Taf Morgannwg and Aneurin Bevan Local Health Boards compared to others (Figure 3).

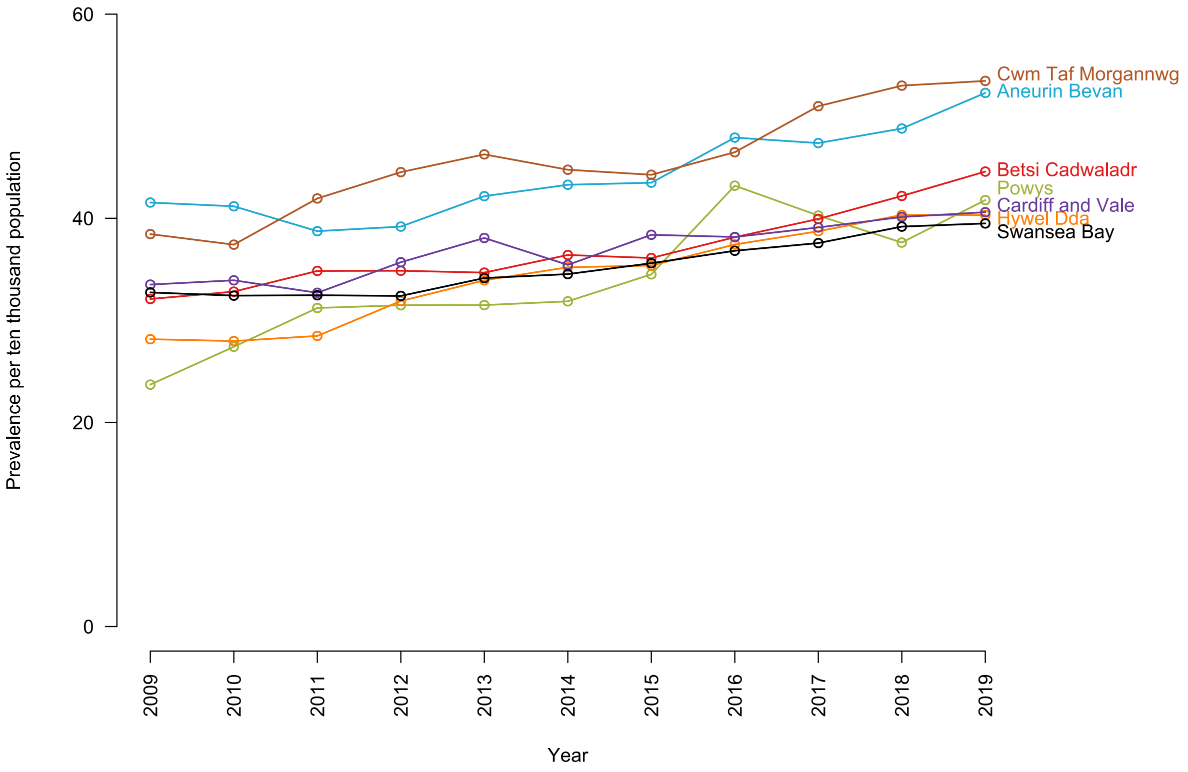


Figure 3: Local Health board variations in prevalence of life-limiting conditions over time.

There were disparities in the distribution of children and young people with life-limiting conditions between the deprivation categories (Figure 4). In contrast to the general population, 20% of whom were in each category, children and young people with life-limiting conditions were more likely to be in the more deprived than the least deprived groups (e.g. in 2019, 25% were in the most deprived group and 18% in the least deprived group).

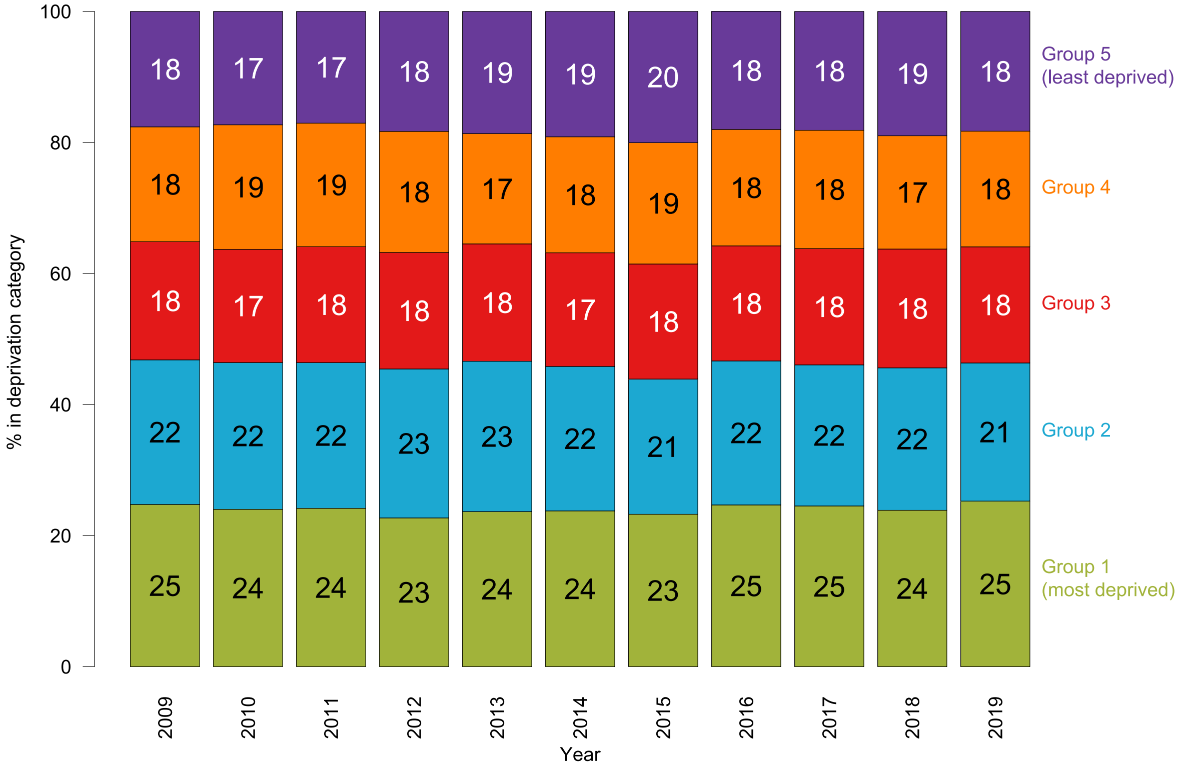


Figure 4: Distribution of children and young people with life-limiting conditions across five deprivation categories. Categories are population-weighted, so if there was an even distribution there would be 20% in each group. The most deprived areas are over- represented. figures on bars indicated the % in each group in each year.

Assessing the prevalence of life-limiting conditions and the proportion of the population that is non-White and the proportion of Lower Super Output Areas in the most deprived category in each Local Health Board, there appears to be some association between deprivation and prevalence (Figure 5). There is no clear relationship between the prevalence of life-limiting conditions and the proportion of Non-White ethnic population but this should be interpreted with caution without individual-level data (Figure 5).

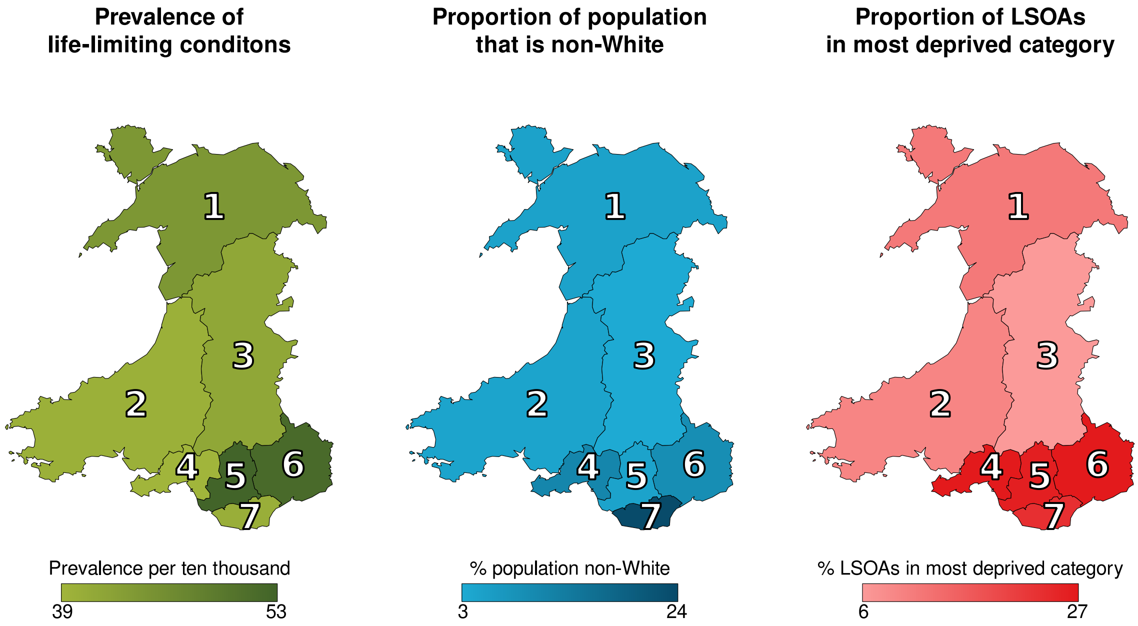


Figure 5: Distributions of prevalence of life-limiting conditions, proportion that is non-White and proportion of LSOAs in the most deprived category across Local Health Boards. There is some correlation between prevalence and Local Health Boards having a greater proportion of the most deprived areas, but no apparent relationship with ethnic group. Local Health Boards: 1. Betsi Cadwaladr UHB; 2. Hywel Dda UHB; 3. Powys THB; 4. Swansea Bay UHB; 5. Cwm Taf Morgannwg UHB; 6. Aneurin Bevan UHB; 7. Cardiff and Vale UHB.

Key Finding 3: Congenital conditions have the highest prevalence, but there are marked increases in prevalence in most diagnostic groups

Overview of methods

Each recorded life-limiting condition was assigned to one of 11 categories of condition - neurology, oncology, metabolic, respiratory, gastrointestinal, haematology, genitourinary, circulatory, perinatal and other. A child or young person with multiple diagnoses could have a life-limiting condition in more than one category. Perinatal diagnoses were only considered for children under 1 year (see appendix for further details).

Results

Congenital conditions had the highest prevalence for children and young people aged under 26 years, three times higher than the next nearest categories, neurology and oncology (2019 prevalence per 10,000: congenital - 24; neurology - 8; oncology - 7) (Figure 6). Most categories of condition saw substantial growth, the only exceptions being respiratory and circulatory conditions with little or no growth over the decade. Metabolic, gastrointestinal and haematology conditions saw the greatest proportional growth over time. Similar distributions were seen for children aged under 18 years.

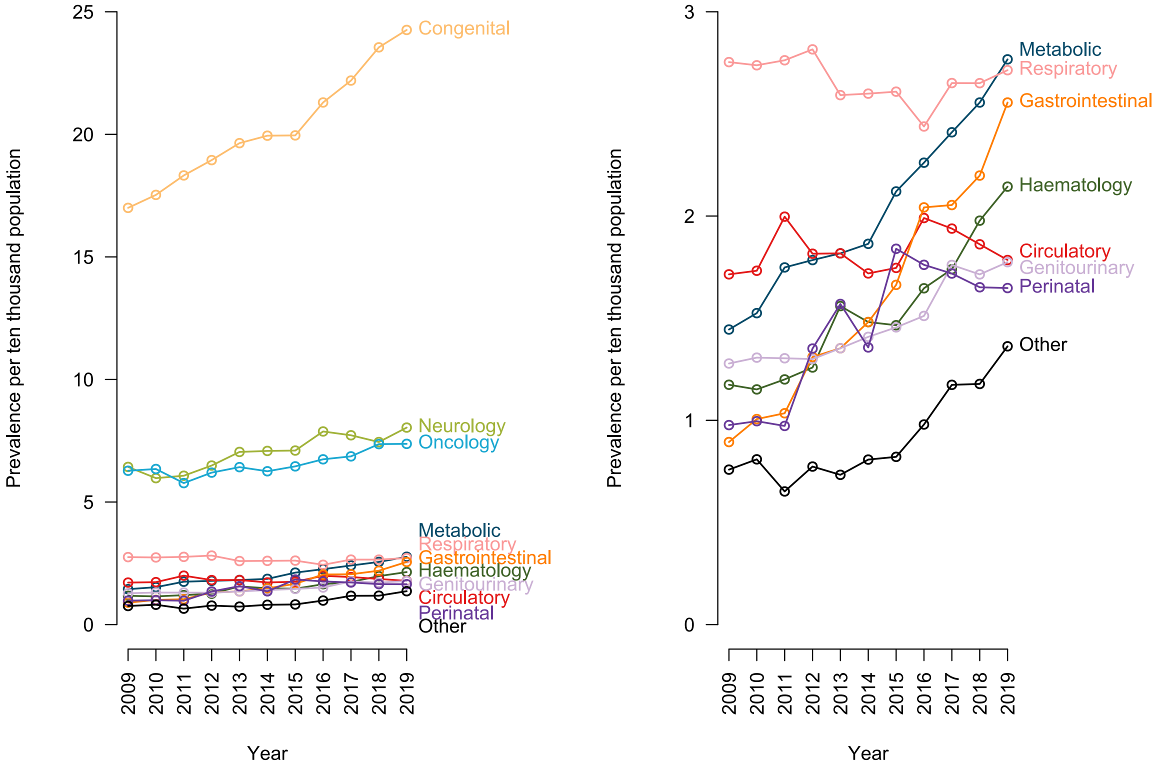


Figure 6: Prevalence, for children and young people aged under 26 years, across the 11 categories of life-limiting condition (left) and detail of prevalence for conditions excluding the three most prevalent (right - excluding congenital, neurology and oncology).

Key Finding 4: Children with life-limiting conditions are disproportionately high users of healthcare

Overview of methods

Data on the use of the following healthcare services were available:

- Inpatient admissions (Wales)
- Paediatric Intensive Care Unit admissions (UK)
- A & E visits (Wales)
- Prescriptions from Primary Care (Wales)

The proportion of healthcare use by children and young people with life-limiting conditions was calculated by first calculating the healthcare use for all children and young people aged 0-17 and 18-25 years from the available data. The same healthcare use for those children and young people with life-limiting conditions was then calculated. Comparing the two enabled quantification of the proportion of health care use for children and young people with life-limiting conditions.

Assessment of prescription use was limited to prescriptions issued in Primary Care, so will underestimate overall prescription use due to missing prescriptions from hospitals and other care settings.(10) It is also important to note that some children with life-limiting conditions have direct ward access so may bypass A&E and we did not have access to data from the NHS in England.

Results

Although a relatively small population in Wales, children and young people with life-limiting conditions have high healthcare use and account for a substantial proportion of hospital and emergency healthcare in Wales.

Children aged under 18 years with life-limiting conditions accounted for 18% of hospital inpatient bed days for children in Wales from 2009-2019 (Figure 7). Within this, they were majority users of paediatric intensive care units (PICUs), accounting for 70% of PICU admissions and 73% of PICU bed days from 2009-2019. They also had disproportionately high numbers of inpatient admissions, emergency inpatient admissions, Accident & Emergency Department visits and General Practice prescriptions.

Figure 8 summarises the disproportionate healthcare use of children with life-limiting conditions (aged under 18 years) in Wales. In 2019, the 1 in 172 children in the population with a life-limiting condition accounted for a majority (7 in 10) of paediatric intensive care unit bed days and 2 in 10 of hospital inpatient bed days. Even the lower proportions of General Practice prescriptions (1 in 14) and Accident & Emergency Department visits (1 in 47) were far in excess of the proportion of children with life-limiting conditions in the population.

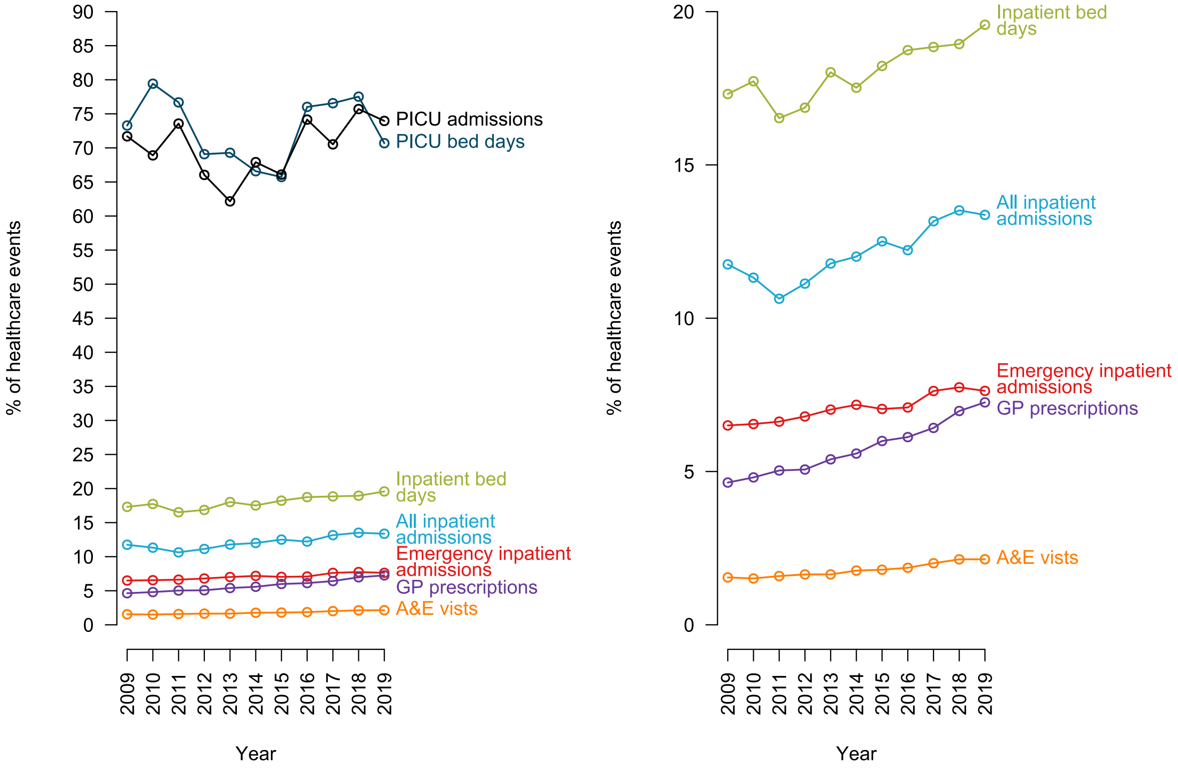


Figure 7: Healthcare use for children and young people with life-limiting conditions diagnosed in childhood as a proportion of healthcare use for all children and young people (left: all healthcare measures; right: detail for healthcare measures excluding paediatric intensive care unit (PICU) admissions and bed days.

Among children:

1 in 172 under 18 years have a life-limiting condition

They account for (among children under 18)**:**

1 in 14 GP prescriptions

1 in 47 A&E visits

2 in 10 hospital inpatient bed days

7 in 10 paediatric intensive care bed days

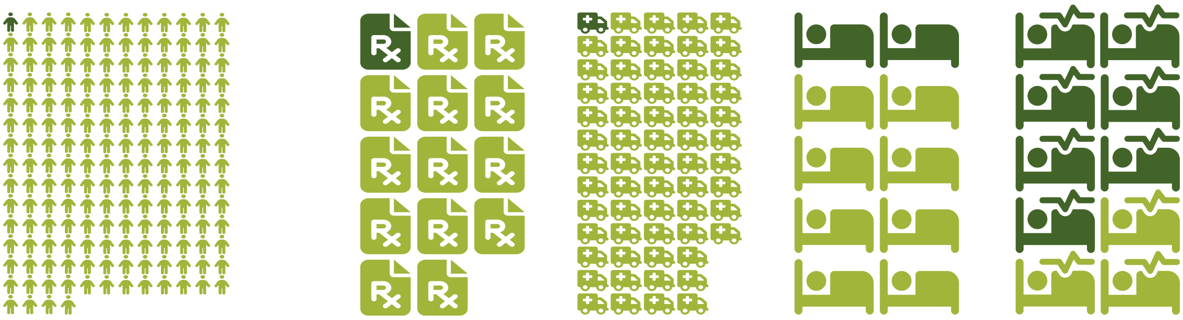


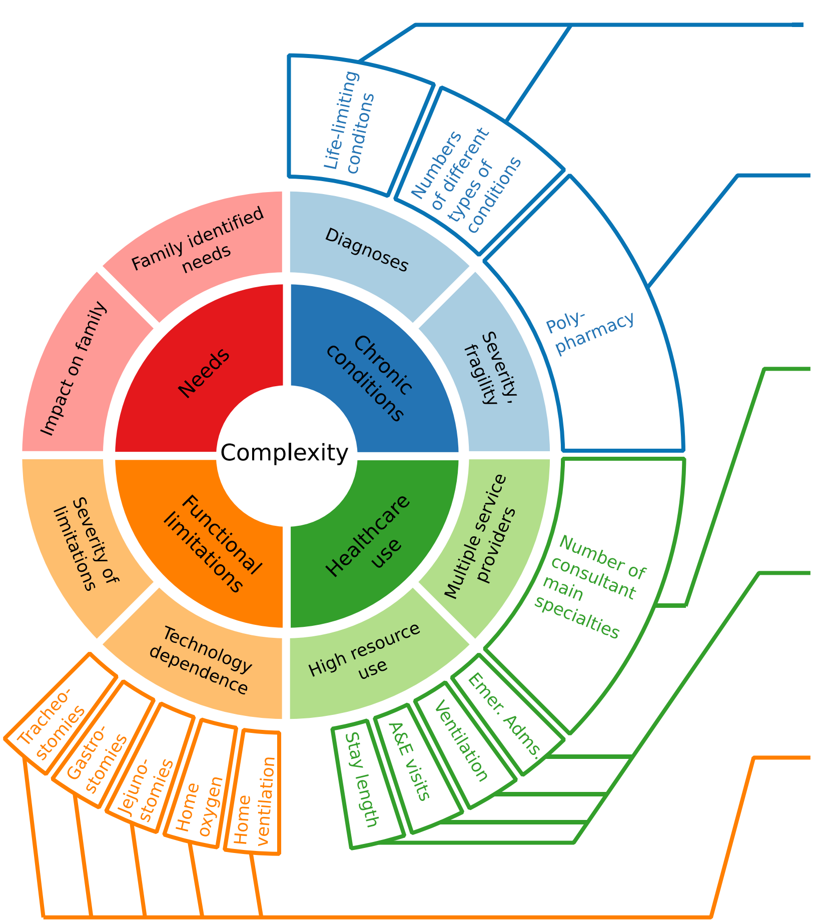
Figure 8: Summary of healthcare use for 2019, putting population size and healthcare use of children (under 18 years) with life-limiting conditions into context compared to all children.

Key Finding 5: There is a range of medical complexity across conditions and increasing complexity with increasing age

Overview of methods

Medical complexity is a multi-dimensional concept that describes how children and young people with life-limiting conditions may need extra time, expertise and resources from healthcare.(11) We used a previous conceptualisation of medical complexity(11) and attempted to quantify this complexity where possible using available healthcare data (Figure 12). We were able to explore aspects of medical complexity relating to diagnoses, severity, use of multiple service providers, high resource use and some aspects of technology dependence. Data were not available to measure the severity of limitations, impact on family or family identified needs.

Each individual scored either zero or one for each of the five components of medical complexity explored (Figure 12). A composite complexity score was then created by adding the component scores, for an overall score from zero to five, with five indicating the highest medical complexity, across each of the five domains. The appendix contains more detail and rationale for the thresholds used.



Score 1 for 'diagnoses' if, recorded over all time, 1 or more of:
- 2+ life-limiting condition categories
- 3+ chronic conditions

Score 1 for 'severity, fragility' if, in year:
- polypharmacy (5+ unique prescriptions) in GP data

Score 1 for 'multiple service providers' if, in year:
- 3+ distinct consultant main specialties recorded in inpatient and outpatient records

Score 1 for 'high resource use' if, in year, any 2 or more of:
- hospital inpatient stay of 10+ days
- 3+ A&E visits
- any ventilation in an intensive care unit
- 2+ emergency inpatient admissions

Score 1 for 'technology dependence' if, in year, any 1 or more of:
- tracheostomy present
- gastrostomoy present
- jejunostomy present
- home oxygen use
- home ventilation

Figure 9: Conceptualisation and scoring system for medical complexity. Complexity is split across four main domains and eight subdomains, five of which can be assessed from items recorded in routine records as indicated. Across each of these five domains, a score of 0 or 1 was assigned based on the criteria indicated; these were then summed to give an overall complexity score between 0 and 5.

Results

The overall complexity score for children and young people (under 26 years) with a life-limiting condition in Wales showed a range of values with all possible scores present, although scores of 1 or 2 were most common (Figure 10) with 25-26% in each. The presence of complexity across all five domains was rare (2%) but 12% of the group could be considered highly complex, scoring 4 or 5. Complexity scores did not differ greatly for children aged under 18 years compared to children and young people aged under 26 years.

There were changes in complexity over time, with a small shift towards higher complexity scores, e.g. in 2009 44% of children and young people with a life-limiting condition had a score of 0 or 1; 10% had a score of 4 or 5. In 2019 the corresponding proportions were 41% and 13%.

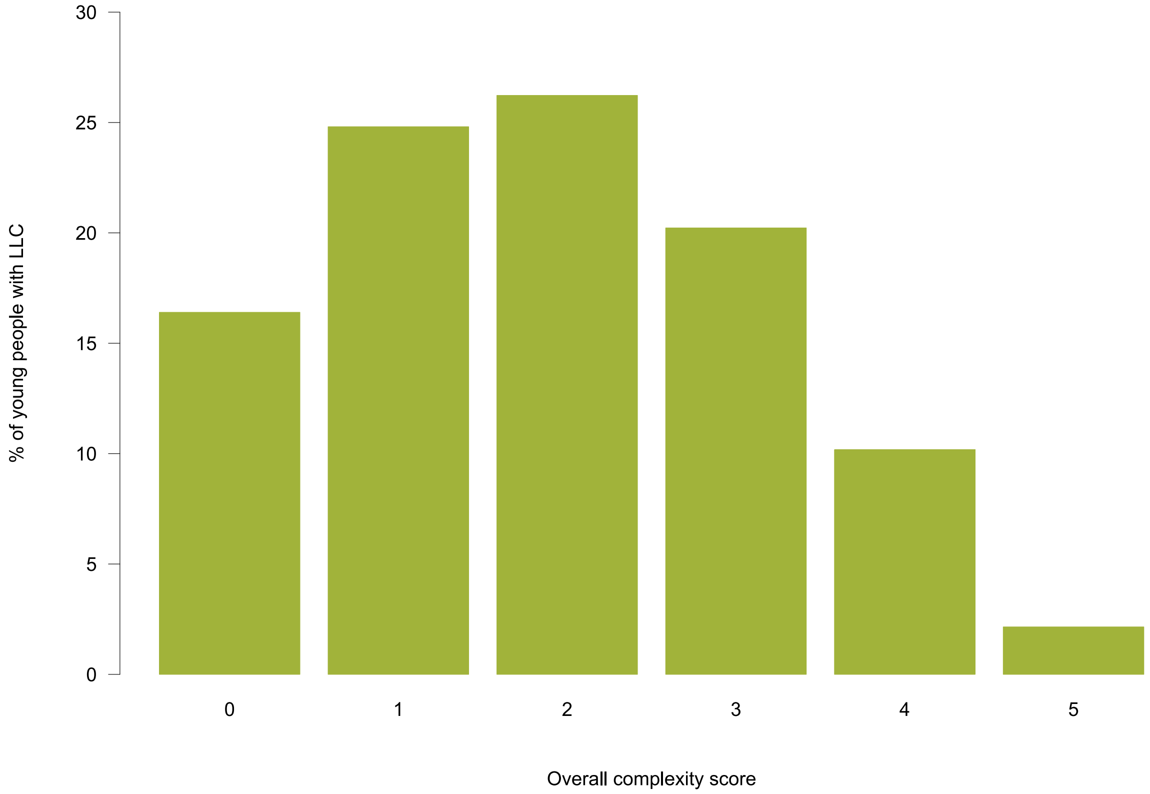


Figure 10: Overall complexity scores for children and young people aged under 26 years with a life-limiting condition diagnosed in childhood in Wales from 2009-2019.

The distribution of complexity scores across ages did differ. Lower scores were much more common in children aged under 1 year than in other age groups (Figure 11). There was a trend towards greater complexity with greater age, which may in part be due to the cumulative nature of some measures - i.e. older children and young people have more time to accumulate more diagnoses than younger children.

Despite the observed differences in prevalence of life-limiting conditions across deprivation categories (Figure 4) there was little variation in complexity across deprivation groups (Figure 12).

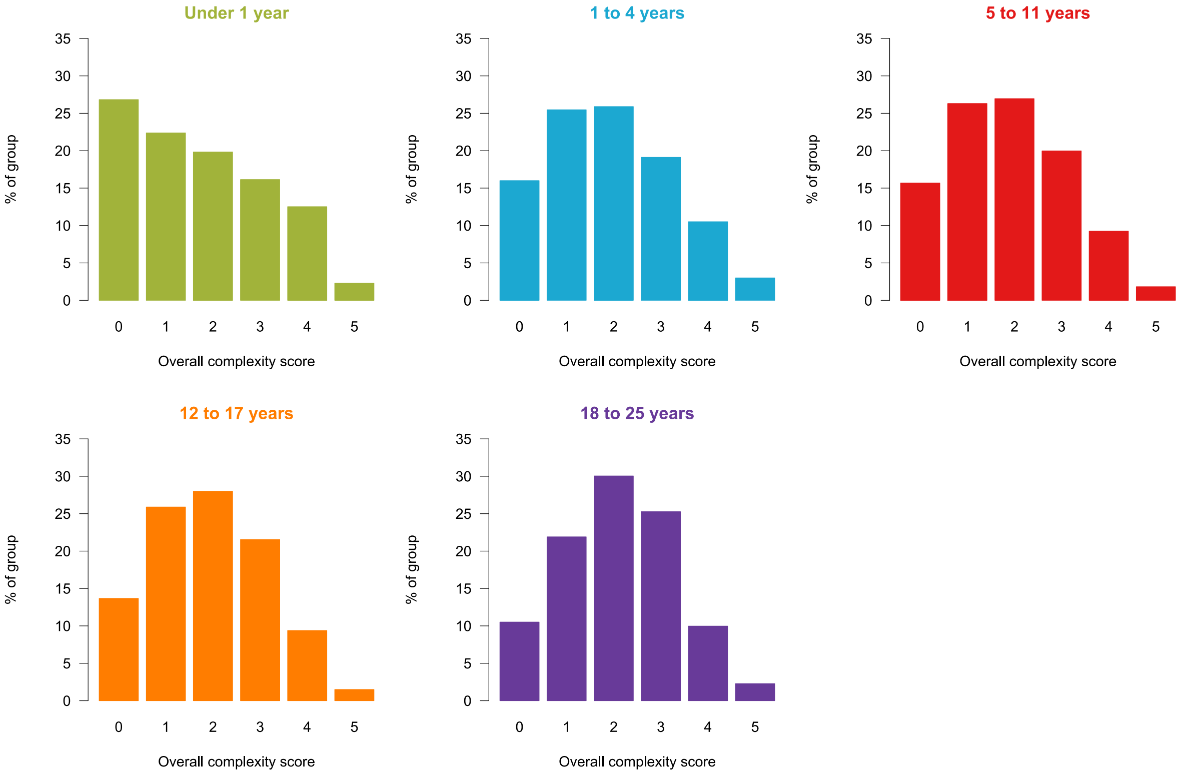


Figure 11: Overall complexity scores by age group for children and young people with a life-limiting condition diagnosed in childhood in Wales from 2009-2019.

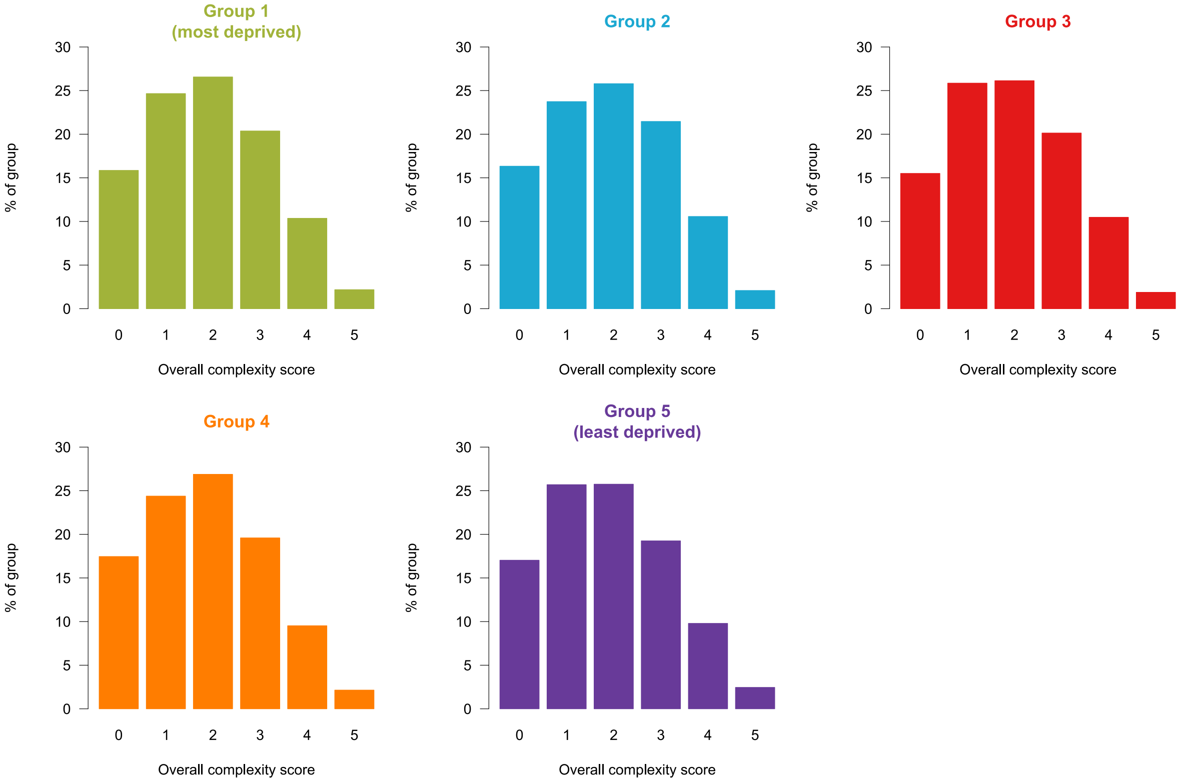


Figure 12: Overall complexity scores by deprivation category for children and young people aged under 26 years with a life-limiting condition diagnosed in childhood in Wales from 2009-2019.

Different distributions of complexity scores were observed across the diagnostic categories of life-limiting conditions (Figure 13). The perinatal category was restricted to children aged under 1 year and the distribution is similar to that observed across all categories of condition for this age group (Figure 11). Respiratory and oncology conditions tended towards lower scores, while conditions classified as 'other' tended towards higher scores, possibly due to the nature of these conditions involving more care teams. The ‘other’ group includes children with rare conditions e.g. graft versus host disease, heart transplant failure or those coded as receiving palliative care. All diagnostic groups had children in the higher levels of complexity.

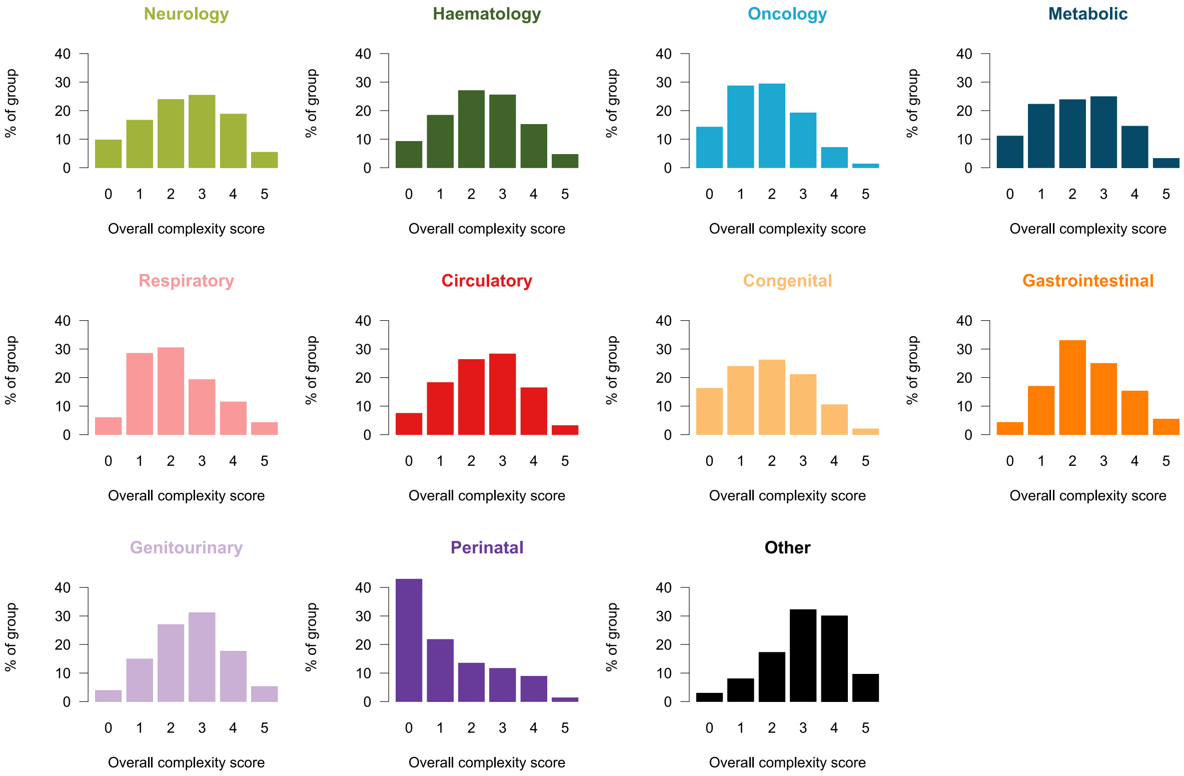


Figure 13: Overall complexity scores by category of condition for children and young people aged under 26 years with a life-limiting condition diagnosed in childhood in Wales from 2009-2019.

Key Finding 6: Approximately a third of children and young people with life-limiting conditions in Wales experience clinical instability each year.

Overview of methods

A previously developed approach to classifying stage of condition (12) was used for this study. Children and young people with life-limiting conditions were assigned one of four stages of condition based on their healthcare use (Figure 14). Children and young people were then classified in each year according to the most severe (highest in Figure 14) stage of condition recorded in that year. Within the available data, very few young people aged 18-25 were in the deteriorating data due to the lack of adult ICU admissions data.

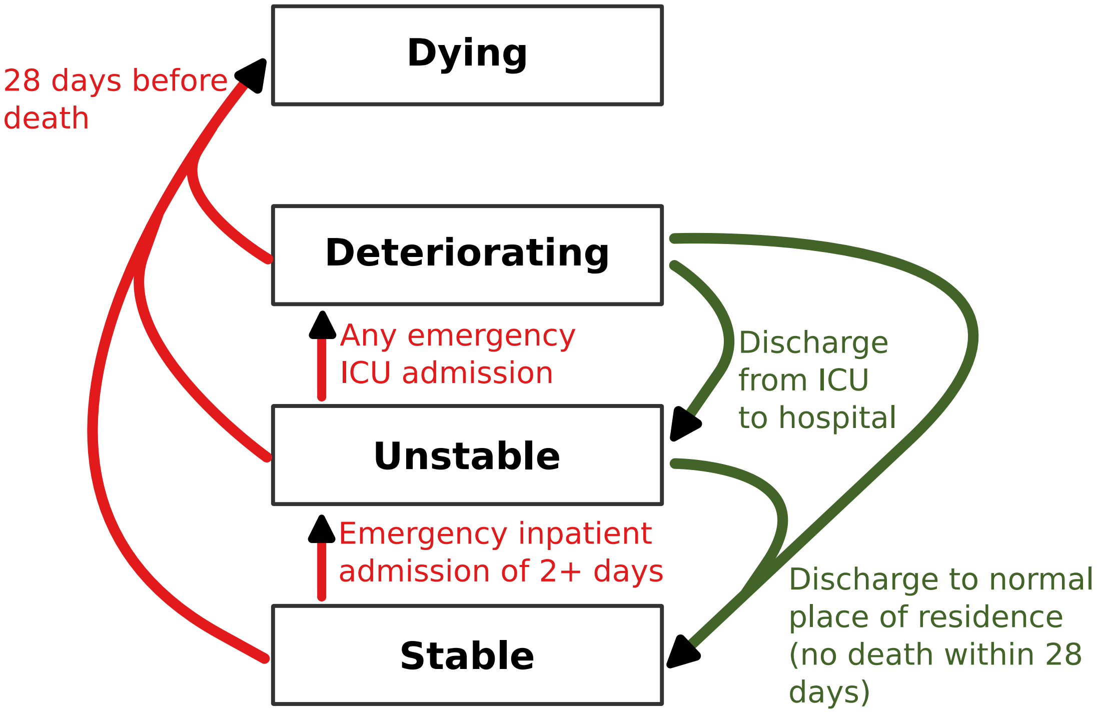


Figure 14: Four stages of condition and the criteria for passing between the stages of condition.

Results

In each year, approximately one third (2019: 32%) of children and young people aged under 26 years with life-limiting conditions experienced clinical instability (i.e. were in a stage of condition other than stable). This varied across the age groups (Figure 15) with, in 2019, 51% of those under 1, 30% aged 1-4 years, 24% aged 5-11 years, 29% aged 12-17 years and 36% aged 18-25 years experiencing clinical instability.

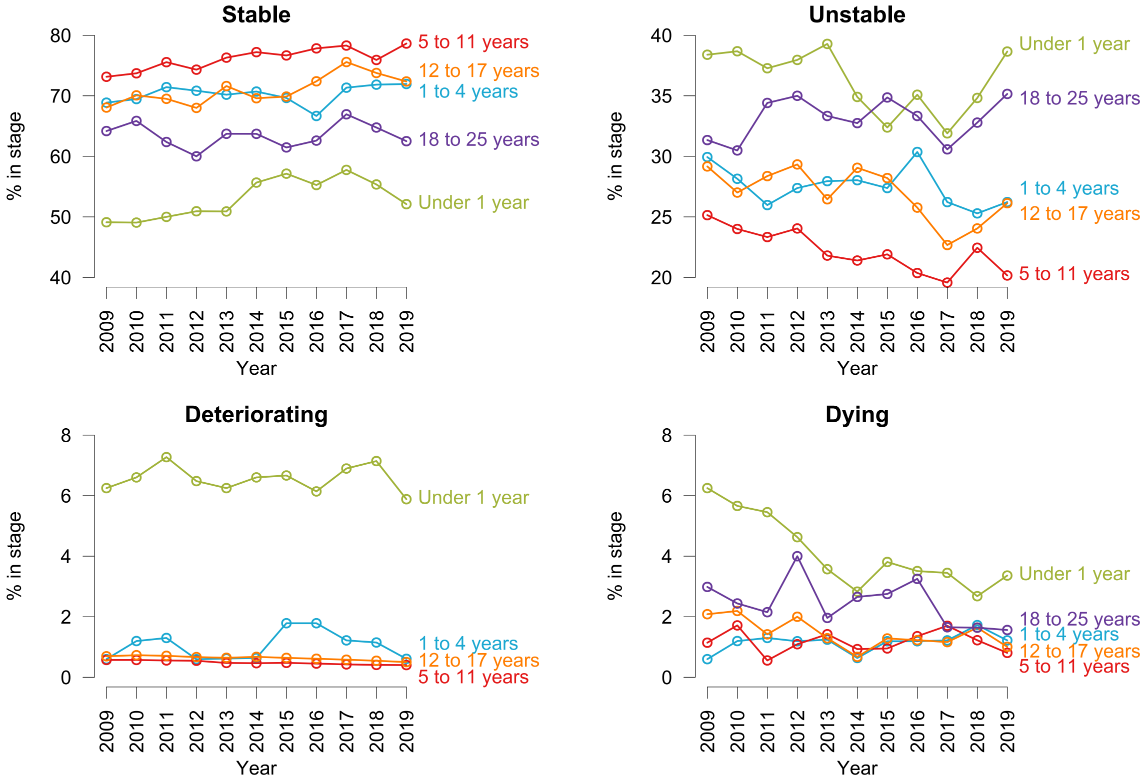


Figure 15: Stage of condition, split by age group, for children and young people in Wales with life-limiting conditions diagnosed in childhood.[3] The 18-25 years age group is omitted from the 'Deteriorating' graph due to a lack of data on intensive care unit admissions for this group.

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[3] The graph for those in the 'Dying' stage of condition in Figure 15 appears to show a substantial fall in the proportion of children under 1 year in this category. Numbers of under 1s dying has remained relatively stable, with only slight decreases, but the number of under 1s with a life-limiting condition diagnosis has increased (there is a corresponding increase in the proportion of under 1s in the 'Stable' stage of condition). These are also quite small numbers, so there is a high degree of variability from year to year which can appear to exaggerate differences.

There were variations between the diagnostic categories of life-limiting condition in the proportion of children and young people experiencing clinical instability in a year (Figure 16). Those with genitourinary conditions were more likely to be stable (51%), while on average 62% in the 'other' category experienced instability each year.

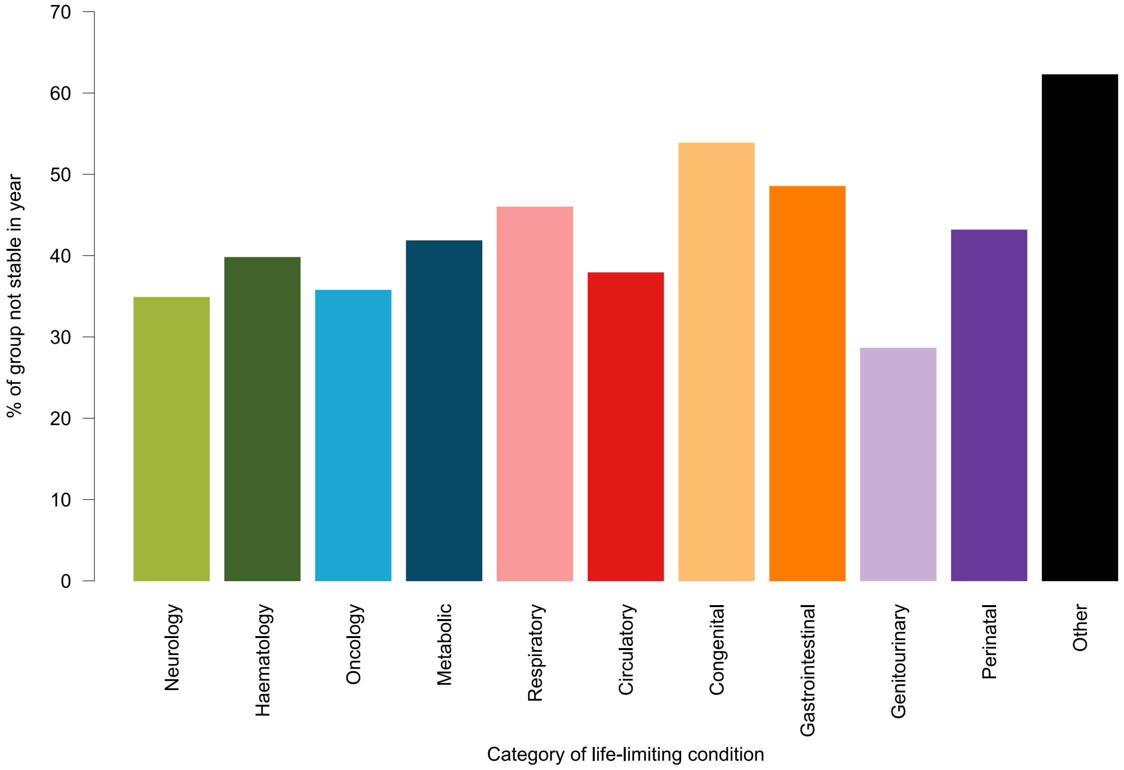

Figure 16: Proportion of children and young people with life-limiting conditions in Wales experiencing at least one period of clinical instability (i.e. unstable, deteriorating or dying) by category of life-limiting condition.

Key Finding 7: Medical complexity and stage of condition are related, but complexity provides additional information on needs

Overview of methods

Complexity scores and stage of condition for each child and young person in each year were compared, to produce distributions of complexity scores by stage of condition. For those in the dying category, complexity scores were recalculated to be for the last year of life, rather than the last calendar year of life as a death early in a calendar year might result in misclassification of complexity status.

Results

Splitting complexity scores by stage of condition reveals that, although there is an association between complexity and stage of condition (stable tends towards lower scores, deteriorating towards higher) there are a range of complexities represented in all stages of condition (Figure 17). This reflects the movement between different stages and that there are few children and young people who can be described as long-term stable or low complexity within this population.

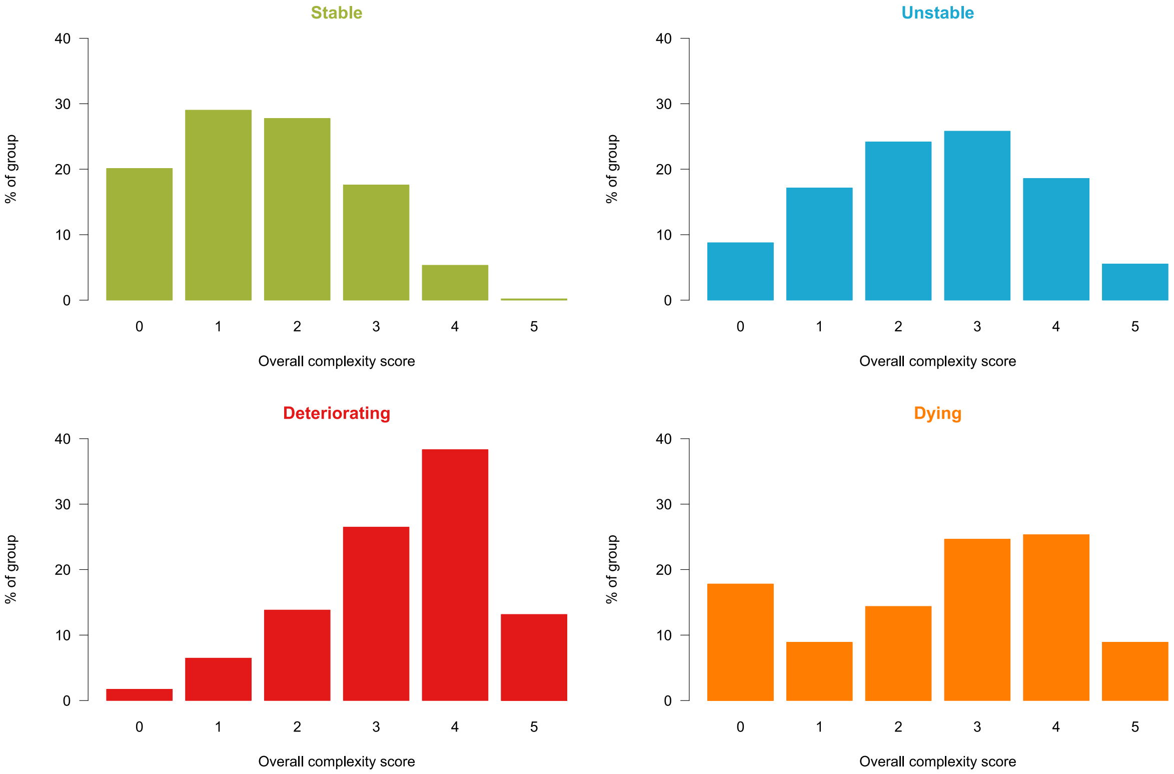

Figure 17: Overall complexity scores, calculated yearly from 2009-2019 (except for those in the category, for whom the score is based on the last year of life) for children and young people with life-limiting conditions in Wales aged under 26 years and split by stage of condition in the year.

Complexity scores in the last year of life (Figure 18) are skewed by the large number of neonatal and infant deaths, with these groups tending towards lower complexity scores. Other age groups have higher scores in the last year of life. This underlines that while the current method of assessing medical complexity is a useful tool for understanding children and young people with life-limiting conditions, it has limitations in picking up complexity in the very young, even in those who subsequently died.

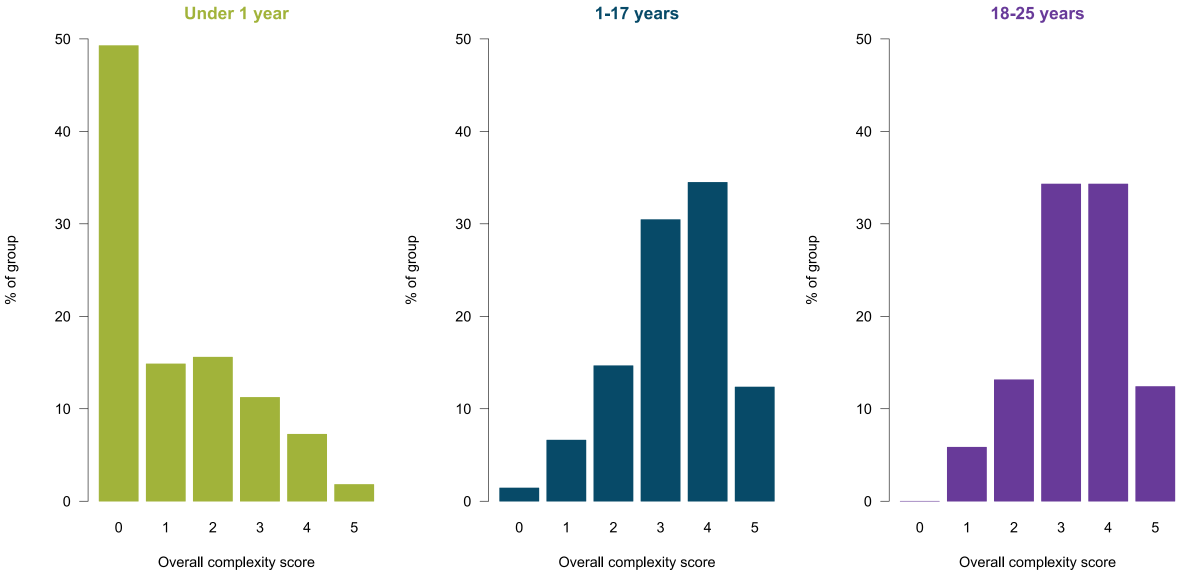

Figure 18: Complexity scores in last year of life by age group, showing the difference between those who died before 1 year and those who died after 1 year.

Key Finding 8: More than half of children who die in Wales have an underlying life-limiting condition

Overview of methods

The Office for National Statistics death registrations were used to identify deaths of Welsh residents aged under 26 years. These were supplemented with reports of deaths from hospital and GP records as there can be a delay in registration of deaths in children(13) (see appendix for details). Children and young people who had died were categorised as either having a life-limiting condition or not having a life-limiting condition, based on the presence of any of the life-limiting condition diagnostic codes on the death certificate or in GP or hospital records before death.

Results

In 2019, 53% (n=105) of children (under 18 years) and 42% (n=145) of children and young people (under 26 years) who died had an underlying life-limiting condition recorded (Figure 19). Overall, from 2009-2019, 49% of children and 40% of children and young people who died had an underlying life-limiting condition recorded. Proportions with a life-limiting condition varied greatly by age with 44% percent in those under 1 year, 27% in those aged 18-25 years and the highest proportions in childhood: 1-4 years - 62%; 5-11 years - 68% (Figure 20). The largest actual number of deaths of those with a life-limiting conditions occur in the under 1 year and 18-25 year age groups (Figure 20).

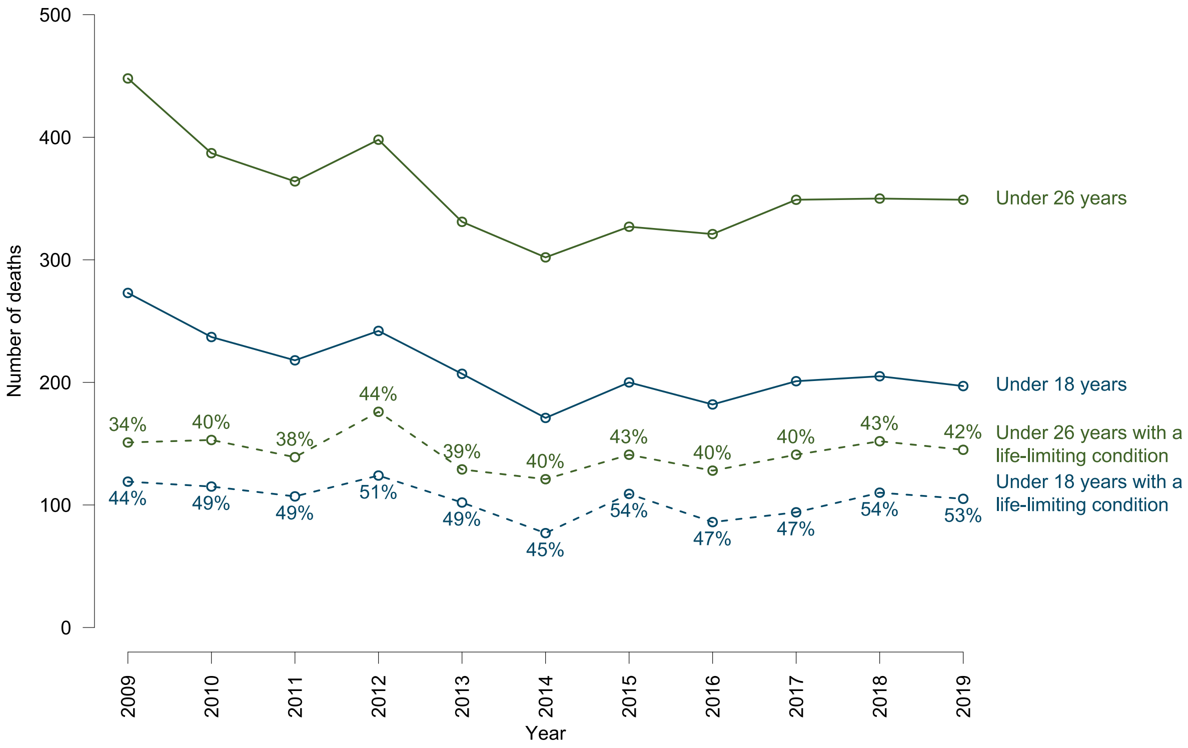

Figure 19: Deaths in Wales from 2009 to 2019 for children (under 18 years) and children and young people (under 26 years) with and without evidence of a life-limiting condition being present. Numbers adjacent to points indicate the proportion of deaths in the year that were for people with a life-limiting condition.

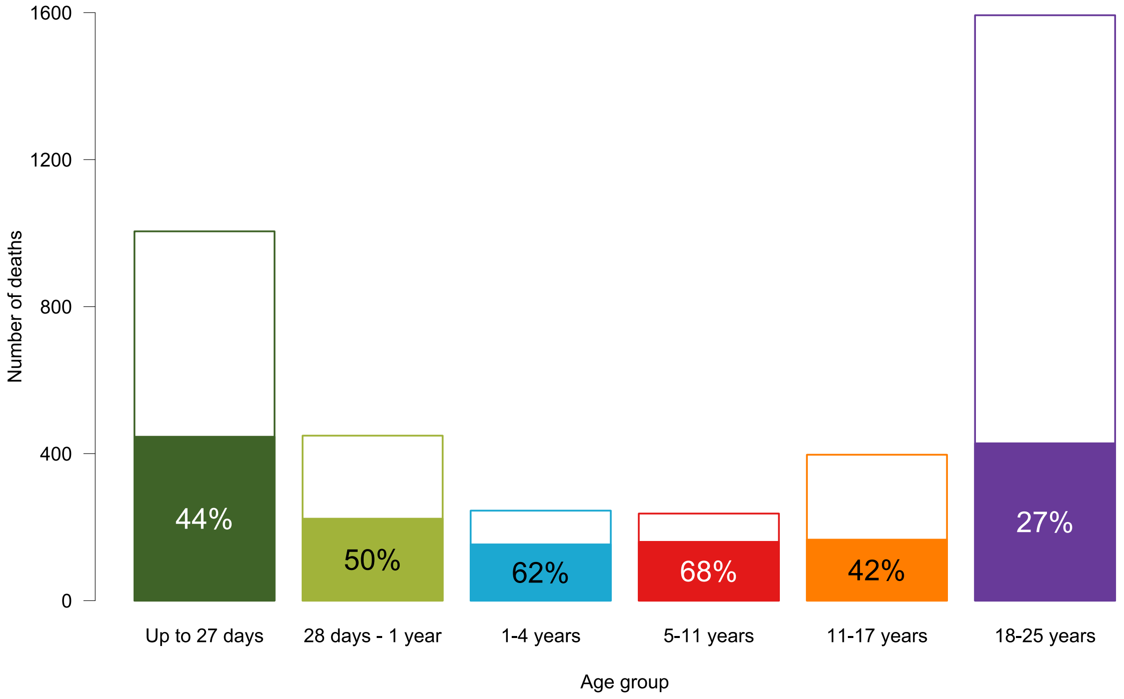

Figure 20: Deaths in Wales by age group with (shaded parts of bars) and without (unshaded parts of bars) a life-limiting condition being recorded as present for 2009-2019. Overall bar height gives the total deaths in that age group for 2009-2019. Numbers on bars are the proportion of deaths in each age group that were for people with life-limiting conditions.

Numbers of deaths, with and without evidence of an underlying life-limiting condition were disproportionately higher in more deprived areas compared to the number of children and young people in each category (20% of the population in each).

Higher numbers of children with underlying life-limiting conditions died in the more deprived areas (Figure 21). However, in less deprived areas a greater proportion of child deaths had evidence of a life-limiting condition compared to more deprived areas (e.g. in the most deprived group, 38% of deaths were for those with a life-limiting condition; least deprived group: 46%). Overall child deaths, apart from those caused by cancer, are related to deprivation.(14)

The highest number of deaths occurred in children with an underlying congenital, oncology or neurological life-limiting diagnoses (Figure 22).

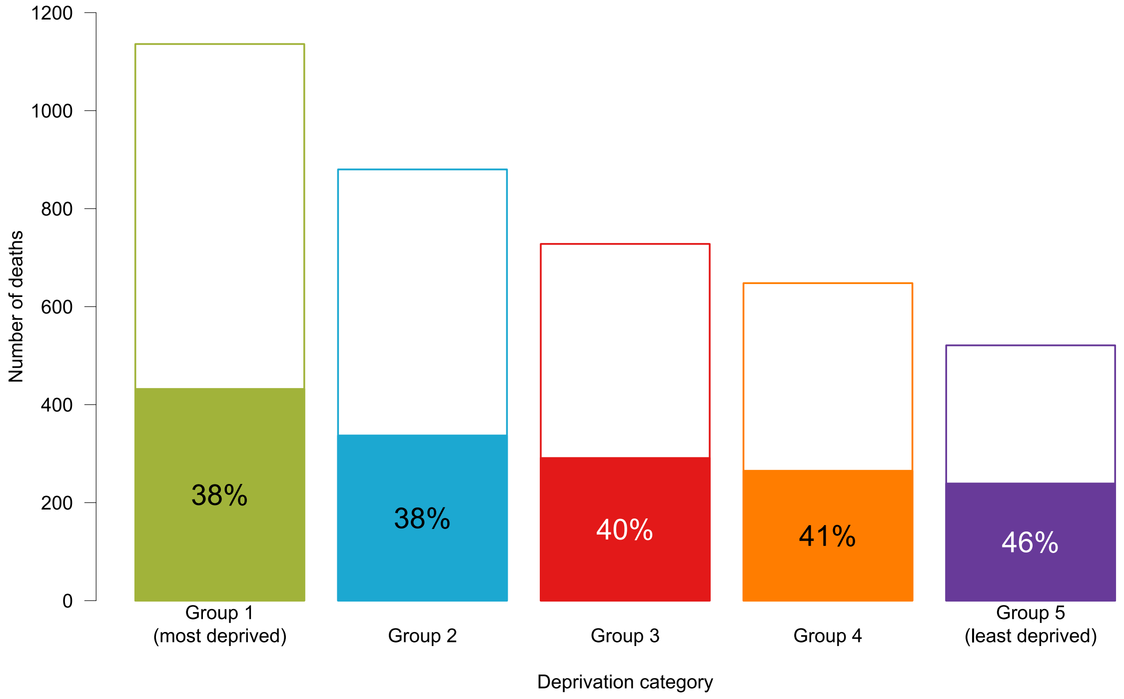

Figure 21: Deaths in Wales for children and young people (aged under 26 years) by deprivation category, with (shaded parts of bars) and without (unshaded parts of bars) a life-limiting condition being recorded as present. Overall bar height gives total deaths in that deprivation category for 2009-2019. Numbers on bars are proportion of deaths within the deprivation category that were for people with a life-limiting condition.

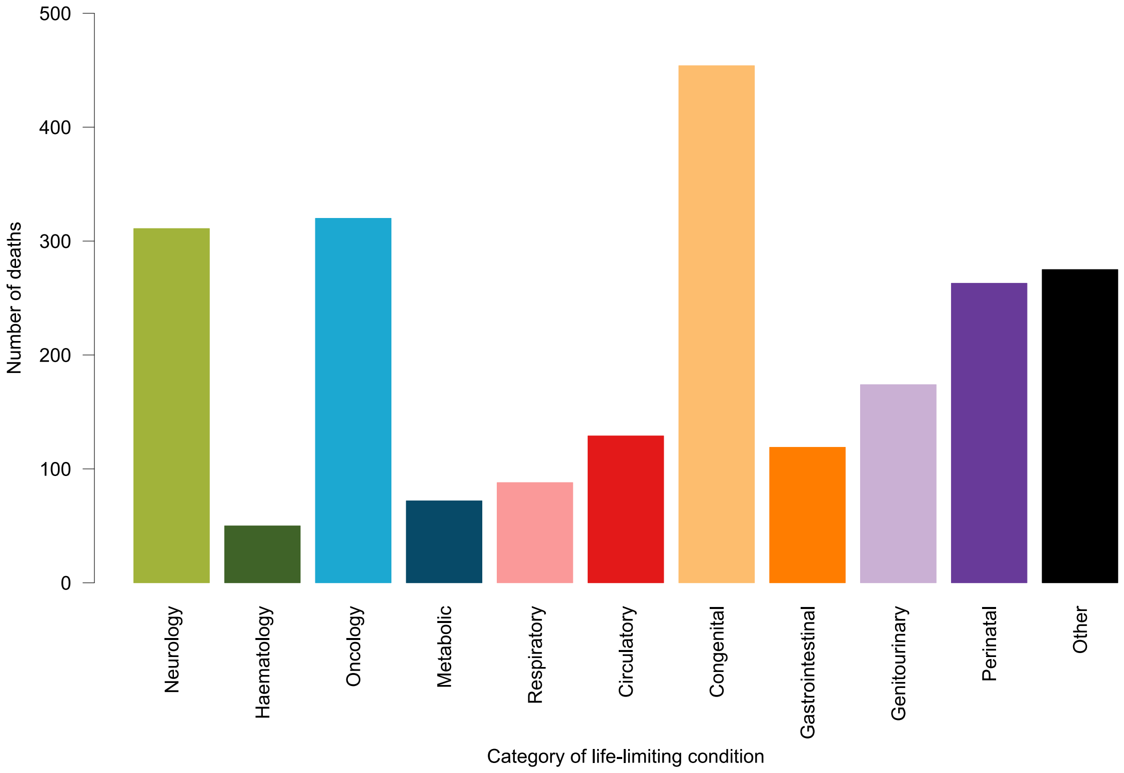

Figure 22: Categories of life-limiting condition recorded for children and young people in Wales known to have died from 2009-2019 (categories are non-exclusive - a death may be recorded under multiple categories).

Strengths and limitations

A strength of this study is its use of national, routinely collected data, ensuring representativeness of the population of Wales. It uses transparent, repeatable methodology (see appendix) and builds on previous work, aiding comparison with past studies and studies in other nations of the United Kingdom.

This study also, for the first time, operationalises a simple measure of medical complexity within routinely collected data. This provides new insights into this population, showing a range of complexity across all diagnostic groups and demographics. By going beyond merely reporting numbers of children and young people with life-limiting conditions, this enables a better understanding of how this population is changing over time and makes it clear that there are few children and young people with life-limiting conditions who can be considered long term stable or of low complexity.

There are also some limitations. The relatively low number of neonates and infants who died with a life-limiting condition identified and the low complexity scores in that group suggest that many life-limiting conditions causing death early in life are missed by current methods and data recording. This is in part due to a lack of specificity in the ICD10 coding framework (i.e. the same diagnosis code may cover a wide range of severity) and the lack of primary care contact for those who die very young (the Read code framework in primary care offers greater specificity). It is also important to note that the measure of complexity here was not able to include severity of limitations, impact on family or family identified needs, due to a lack of recording of this in the data. This is an important limitation and consideration should be given to ways in which these data might be captured in future.

There were other limitations due to available data. The inability to obtain hospital data from England (other than for paediatric ICU admissions from PICANet) means that some healthcare events and potentially some children and young people with life-limiting conditions will have been missed, particularly if most of their care takes place in England. This cannot be quantified. Future studies would benefit from availability of these data, or gathering of this data within Wales for research purposes (these data are passed between England and Wales for payment purposes).

The poor quality and incompleteness of ethnic group recording in the available datasets prevented disaggregation of results by ethnic group. Given previous findings of a relationship between prevalence of life-limiting conditions and ethnic group, these data would help to ensure targeting and cultural appropriateness of services where needed. Improvement of gathering of these data should be a priority.

Primary care data are limited for young children with life-limiting conditions, who may not register with or see a GP while under hospital care. Some data items are also limited or incomplete for this population in primary care - i.e. prescriptions, where many prescriptions may originate from hospitals or other community services. There were also very few instances of diagnoses or prescriptions relating to home ventilation, suggesting this is either poorly recorded or comes from other sources.

We lacked data on adult ICU admissions in the analysed datasets, limiting analyses on stage of condition and healthcare use for young people aged 18-25 years. There will be underestimates of severity of stage of condition and healthcare use in this group.

Finally, whilst the strengths of these data are the full population data sources available, improvements to data collection and quality should be considered. No data from children’s hospices or paediatric palliative care providers could be included in these analyses as these data are not routinely linked to other healthcare data in Wales. The lack of child or parent reported outcome measures limits future analyses of quality of care.

Concluding Comments

The aim of this report was to summarise the key findings on the population of children and young people in Wales with a life-limiting or life-threatening condition. This included identifying the numbers of children, prevalence by age, deprivation and health board and complexity of needs of this population of children and young people.

An analysis of routinely collected healthcare data was undertaken to provide this information. Wales has access to full population linked primary and secondary care data, but we did not have access to data for those who had access the NHS in England, therefore there may be children missing from these analyses. This report was also limited by a lack of ability to link data from children hospice and paediatric palliative care services.

The increasing numbers and prevalence of this population is similar to patterns seen in other parts of the UK (4, 15, 16) and is likely to be driven by increasing survival in this population.(15)

The availability of full population linked primary and secondary healthcare data has enabled us to estimate levels of complexity of needs in this population for the first time. The results show a range of complexities but also highlight that these measures may be less helpful in assessing complexity in the under 1 age group.

The population of young people aged 18-25 years who were diagnosed with a life-limiting condition in childhood were included in this report. They are also a growing population, have high levels of complexity and a number of these young people will require palliative and end-of-life care every year.

All these findings are vital for the policy makers, healthcare planners and service providers to inform healthcare provision for this population of children and young people. The disparities reported here in terms of age, deprivation and health board are very important if accessible and equitable care is to be provided for this population of children and young people.

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Melanie Lewis (NHS Wales Health Collaborative)

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Appendix: Detailed methods

Definition of life-limiting and life-threatening conditions

Life Limiting conditions are those for which there is no reasonable hope of cure and from which children or young people will die. Life-threatening conditions: are those for which curative treatment may be feasible but can fail, such as cancer.(6)

This study built upon previously developed and refined lists of life-limiting condition diagnoses,(15, 16) using diagnostic codes from primary (Read code) and secondary (International Classification of Diseases, 10th Edition(17) - ICD10) care.

Diagnoses in hospital records

The development of the ICD10 coding framework has been previously described.(16) The full list of ICD10 codes is provided in Table A 1. In common with a previous study in England (15) those conditions categorised as perinatal (see table A1) were only included for children aged under 1 year - children and young people aged 1 year and over were only considered to have a life-limiting condition if they had a life-limiting diagnosis in one of the other categories of condition. This was to prevent long term inclusion of children and young people who may have had a life-threatening event around the time of birth but did not have a life-limiting condition in the longer term. In contrast to the previous study (15) we did not restrict the time for which, from an original oncology diagnosis, a child or young person could be considered to have a life-limiting condition. This was due to the observed small effect of this restriction in the previous study, the requirement in the present study for ongoing inpatient admissions for a child or young person to be included (i.e. a young person with childhood cancer, now cured and no longer requiring inpatient care would not be included) and the risk of excluding recurring or new oncology diagnoses under this restriction.

Diagnoses in primary care records

The Read code diagnosis framework for use with primary care records was initially developed through manual translation of diagnoses into the Read system.(18) For this study, this list was further refined and expanded by translating the ICD10 coding framework into Read Codes using the *CALIBERcodelists* package[4] for the *R*[5] statistical computing environment; the generated Read codes were then screened for inclusion or exclusion. The full list is available on request.

Population data

The size of the population in Wales within the age group(s) studied within Local Health Boards was derived from census 2021 data from StatsWales[6]. Data on ethnic groups by Local Health Board were obtained from the NOMIS website[7]. Data on distributions of deprivation categories by Local Health Board were obtained from StatsWales[8].

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[4] https://rdrr.io/rforge/CALIBERcodelists/

[5] https://r-project.org

[6] https://statswales.gov.wales/Catalogue/Population-and-Migration/Population/Estimates/Local-Health-Boards/populationestimates-by-lhb-age

[7] https://www.nomisweb.co.uk/datasets/c2021rm032

[8] https://statswales.gov.wales/Catalogue/Community-Safety-and-Social-Inclusion/Welsh-Index-of-Multiple-Deprivation/WIMD-2019/localhealthboardanalysis

Table A 1: List of ICD10 codes, arranged by diagnostic group, used to identify life-limiting conditions in secondary care data and death records.

Neurology	Haematology	Oncology	Metabolic	Respiratory	Circulatory	Gastrointestinal	Genitourinary	Perinatal	Congenital		Other
A17	B20-B24	C00-	E31.0	E84	I21	K55.0	N17	P10.1	Q00.0	Q44.2	H11.1
A81.0	D56.1	C97	E34.8	J84.1	I27.0	K55.9	(exlc	P11.2	Q01	Q44.5	H49.8
A81.1	D61.0	D33	E70.2	J96	I42	K72	N179)	P21.0	Q03.1	Q44.7	H35.5
F80.3	D61.9	D43	E71	(excl	I61.3	K74	N18	P28.5	Q03.9	Q60.1	M31.3
F84.2	D76.1	D44.4	E72	J96.9)	I81	K76.5	(excl	P29.0	Q04.0	Q60.6	M32.1
G10	D81	D48	E74	J98.4		K86.8	N181,	P29.3	Q04.2	Q61.4	M89.5
G11.1	D82.1		E75				N182,	P35.0	Q04.3	Q61.9	T86.0
G11.3	D83		E76				N183)	P35.1	Q04.4	Q64.2	T86.2
G12	D89.1		E77				N19	P35.8	Q04.6	Q74.3	Z51.5
G20			E79.1				N25.8	P37.1	Q04.9	Q74.8	
G23.0			E83.0					P52.4	Q07.0	Q75.0	
G23.8			E88.0					P52.5	Q20.0	Q77.2	
G31.8			E88.1					P52.9	Q20.3	Q77.3	
G31.9								P83.2	Q20.4	Q77.4	
G35								P91.2	Q20.6	Q78.0	
G40.4								P91.6	Q20.8	Q78.5	
G40.5								P96.0	Q21.3	Q79.2	
G60.0									Q23.2	Q79.3	
G60.1									Q21.8	Q80.4	
G70.2									Q22.0	Q81	
G70.9									Q22.1	Q82.1	
G71.0									Q22.4	Q82.4	
G71.1									Q22.5	Q85.8	
G71.2									Q22.6	Q86.0	
G71.3									Q23.0	Q87.0	
G80.0									Q23.4	Q87.1	
G80.8									Q23.9	Q87.2	
G82.3									Q25.4	Q87.8	
G82.4									Q25.6	Q91	
G82.5									Q26.2	Q92.0	
G93.4									Q26.4	Q92.1	
G93.6									Q26.8	Q92.4	
G93.7									Q28.2	Q92.7	
									Q32.1	Q92.8	
									Q33.6	Q93.2	
									Q39.6	Q93.3	
									Q41.0	Q93.4	
									Q41.9	Q93.5	
									Q43.7	Q93.8	
										Q95.2	

Datasets used and data management

SAIL Datasets

All data analysis was conducted within the SAIL Databank,(5) one of the UK's Secure eResearch Platforms, providing a secure environment for the analysis. SAIL standard datasets used are summarised in Table A 2.

Table A 2: SAIL standard datasets used in this study, including purpose of use.

Dataset (short names in SAIL)	Use in this study
Annual District Death Extract (ADDE)	• Stage of condition - entering dying category • Deaths - date of death, cause of death
Congenital Anomaly Register and Information Service (CARIS)	• Deaths - date of death
Emergency Department Data Set (EDDS)	• Healthcare use - A&E visits • Complexity - A&E visits • Deaths - date of death • Demographic data
Outpatient Database for Wales (OPDW)	• Complexity - number of main consultant specialties • Demographic data
Patient Episode Database for Wales (PEDW)	• Identification of life-limiting conditions • Presence in Wales with inpatient admission • Healthcare use - admissions, emergency admissions, bed days • Complexity - emergency admissions, length of stay • Stage of condition - emergency admissions • Demographic data
Wales Longitudinal General Practice Dataset (WLGP)	• Identification of life-limiting conditions • Complexity - GP prescriptions

Data from the Paediatric Intensive Care Audit Network (PICANet)

In addition to the standard SAIL datasets, data were requested from PICANet[9] on UK-wide paediatric intensive care unit admissions for residents of Wales aged under 26 years. These were then linked by SAIL with the standard SAIL datasets.

Data from NHS Digital

It was intended to also use data on England hospital care for Wales residents aged under 26 years. This was to ensure inclusion of children and young people and healthcare use taking place outwith Wales (e.g. for some Wales residents the nearest major hospital may be in England or some specialist treatments may only be available at centres in England). Unfortunately, NHS Digital were unable to provide these data in time for inclusion in this study.

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[9] https://picanet.org.uk/

Data management

Presence in Wales with an inpatient admission

Children and young people were only included in the analyses in each year (with the exception of the analyses of deaths) if they had an inpatient admission, while resident in Wales and aged under 26 years. This was determined from the PEDW datasets.

Demographic variables

Demographic data were determined from the data as follows:

Age

Age in each year was set to age at the first hospital inpatient admission in the year (to be included in the analyses, children and young people were required to have a hospital admission in the year)

Sex

Sex was set to the most commonly recorded, from all available records in the hospital inpatient (PEDW), hospital outpatient (OPDW) and Emergency Department (EDDS) records.

Ethnic group

Ethnic group data were gathered from PICANet data, hospital inpatient (PEDW), hospital outpatient (OPDW) and Emergency Department (EDDS) records. Recorded ethnic groups were combined into seven main groups, based on census categories: Bangladeshi, Black, Chinese, Indian, Pakistani, White and Mixed/Other. The most commonly recorded of these seven categories was then assigned. Unfortunately, large quantities of missing data prevented these categories from being used in these analyses.

Local Health Board

Local Health Board was assigned based on Lower Super Output Area (2011 - a small unit area of geography, each with about 1500 people[10]) of residence from hospital inpatient (PEDW), hospital outpatient (OPDW) and Emergency Department (EDDS) records. The first non-missing Wales LSOA was used in each year. These were mapped to Local Health Board using mappings from the Office for National Statistics[11].

Deprivation category

LSOA (2011) values were determined as described above. These were mapped to Welsh Index of Multiple Deprivation (WIMD) 2019 scores using data from The Welsh Government[12] and then ordered by score and split into five equal population (for the under 26 years population) groups.

Diagnoses

Life-limiting condition diagnoses were identified from hospital care by combining the relevant hospital inpatient admissions datasets (PEDW) and matching recorded diagnoses from all available fields against the ICD10 coding framework. Each matching code was classified into one of the eleven categories of condition and the date recorded.

Life-limiting condition diagnoses were identified from primary care by matching diagnostic events from primary care records (WLGP) against the Read code coding framework. Each matching code was classified into one of the eleven categories of condition and the date recorded.

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[10] https://www.gov.wales/docs/statistics/lsoamaps/lsoa.htm

[11] https://geoportal.statistics.gov.uk/datasets/efdb99e8351a42878700f08e6d8787d7_0/explore

[12] https://www.gov.wales/welsh-index-multiple-deprivation-full-index-update-ranks-2019

These relevant diagnoses were then combined and, for each person, the first date of diagnosis with a life-limiting condition, the first date of diagnosis with a non-perinatal life-limiting condition and the first date of diagnosis (if any) in each of the categories of life-limiting condition was recorded.

Analyses

Numbers and prevalence

Numbers of children and young people with life-limiting conditions were determined in each year by counting those who met all the following requirements:

- Aged 0-25 in year
- With an inpatient admission in the year while resident in Wales
- One of:
    - First date of life-limiting condition (excluding perinatal conditions) was in year or an earlier year AND before age 18 years

    OR

    - First date of perinatal life-limiting condition was recorded in year or a previous year AND age in year was 0

These numbers were then disaggregated by age, sex, Local Health Board and deprivation category, using the derived demographic data (see details above).

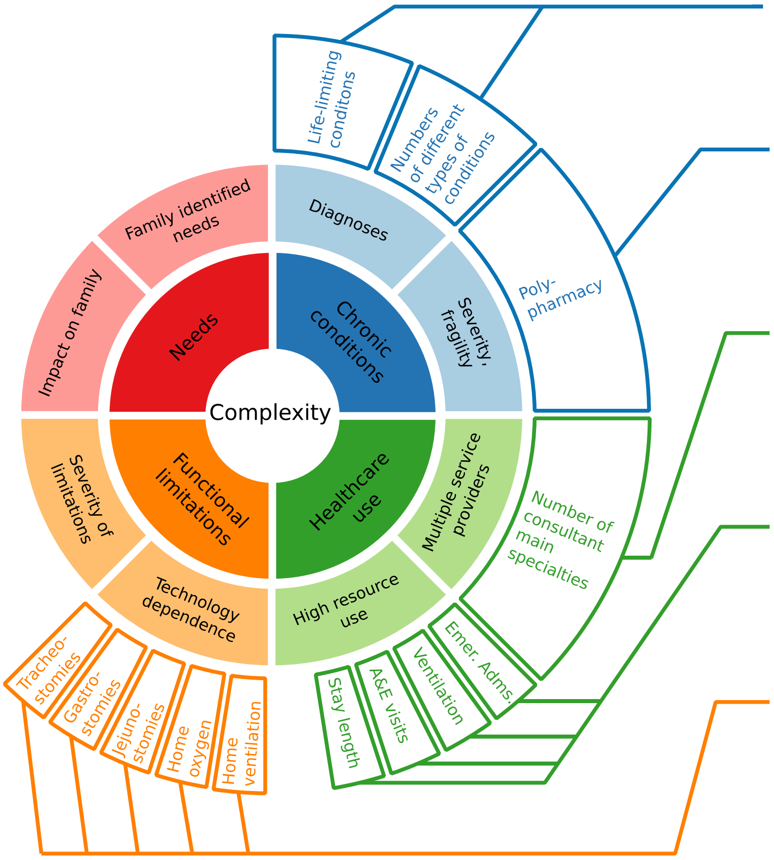
Healthcare use

Healthcare use events were counted for the whole population of Wales residents and for those determined (see preceding section) to have a life-limiting condition in the year, as follows:

- General Practice prescriptions: unique prescriptions recorded in GP data (WLGP)
- A&E visits: all events with arrival date in Accident & Emergency datasets (EDDS)
- All inpatient admissions: all admissions in inpatient data (PEDW)
- Emergency inpatient admissions: admissions in inpatient data (PEDW) where admission method (ADMIS_MTHD_CD) began with '2' and was not '2C' (Emergency admission: baby born at home as intended)
- Inpatient bed days: all admissions in inpatient data (PEDW) with bed days equal to the difference between START_DATE and END_DATE plus 1 (i.e. an admissions and discharge on the same day is 1 bed day)
- Paediatric intensive care unit admissions: All admissions in PICANet data
- Paediatric intensive care unit bed days: All admissions in PICANet data, with bed days equal to the difference between ADDATE and UNITDISDATE plus 1 (i.e. an admissions and discharge on the same day is 1 bed day)

Medical complexity

Medical complexity scoring built upon previous conceptualisation work (11) and initial attempts to operationalise this in routinely collected healthcare data in England.(19) Scores were summed over five domains, with each domain being given a score of 0 or 1 (Figure A 1) giving an overall score from 0 to 5.



Score 1 for 'diagnoses' if, recorded over all time, 1 or more of:
- 2+ life-limiting condition categories
- 3+ chronic conditions

Score 1 for 'severity, fragility' if, in year:
- polypharmacy (5+ unique prescriptions) in GP data

Score 1 for 'multiple service providers' if, in year:
- 3+ distinct consultant main specialties recorded in inpatient and outpatient records

Score 1 for 'high resource use' if, in year, any 2 or more of:
- hospital inpatient stay of 10+ days
- 3+ A&E visits
- any ventilation in an intensive care unit
- 2+ emergency inpatient admissions

Score 1 for 'technology dependence' if, in year, any 1 or more of:
- tracheostomy present
- gastrostomoy present
- jejunostomy present
- home oxygen use
- home ventilation

Figure A 1: Conceptualisation and scoring system for medical complexity. Complexity is split across four main domains and eight subdomains, five of which can be assessed from items recorded in routine records as indicated. Across each of these five domains, a score of 0 or 1 was assigned based on the criteria indicated; these were then summed to give an overall complexity score between 0 and 5.

The rationale for the scoring criteria was as follows:

- Diagnoses: these (either of two or more life-limiting condition categories OR 3 or more chronic conditions) were chosen from distributions in the data, setting thresholds that would give a range of complexity scores within this population.
- Polypharmacy: five or more unique prescriptions is a previously used definition of polypharmacy.(10)
- Multiple service providers: 3 or more unique consultant main specialities was chosen by looking at distributions in the data, setting thresholds that would give a range of complexity scores within this population.
- High resource use: given the relatedness between measures, it was chosen to require two or more of:
  - hospital stay of 10 or more days (previously identified as a long stay in this population)(20)
  - three or more A&E visits (approximately 4 times the number expected across this age group for children with life-limiting conditions)(18)

    - two or more emergency admissions (approximately 4 times the number expected across this age group for children with life-limiting conditions)(18)
    - ventilation in paediatric intensive care unit: considered a marker of severity.
- Technology dependence: gastrostomy, tracheostomy, jejunostomy presence, use of home oxygen or home ventilation were all considered dependent on technology.

These were measured as follows:

- Diagnoses: life-limiting conditions were assessed using categories identified as described above. Chronic conditions were assessed by matching a previously developed list of chronic conditions(21) to inpatient diagnoses (PEDW). These were limited to inpatient diagnoses to restrict to conditions meriting or considered relevant to inpatient care.
- Polypharmacy: as described above for healthcare use.
- Multiple service providers: unique consultant main specialties in each year were counted from inpatient (PEDW) and outpatient (OPDW) datasets.
- High resource use: as described above for healthcare use; ICU ventilation was identified from PICANet data by completion of any of the HASINVVENTTDAYS, HASINVENTTTTDAYS, HASNIVDAYS, HASAVSJETDAYS or HASAVSOSCDAYS fields.
- Technology dependence: for gastrostomies, tracheostomies and jejunostomies, relevant ICD10, OPCS4 and Read codes[13] were searched for in inpatient (PEDW) and primary care (WLGP) records. For home ventilation and home oxygen use, relevant diagnostic and prescription codes were searched for in primary care data (WLGP).

Stage of condition

Stages of condition were defined as set out in Figure A 2, following previous work.(12)

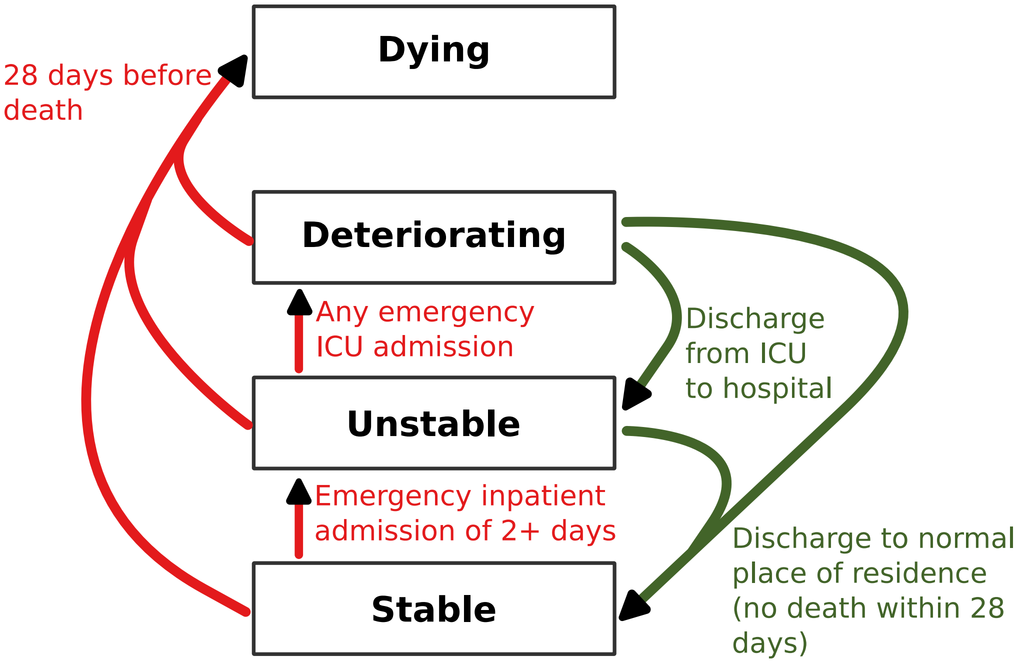


Figure A 2: Four stages of condition and the criteria for passing between the stages of condition.

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[13] ICD10, OPCS4 and Read code lists available on request

Emergency inpatient admissions were identified as set out above in the Healthcare Use section. Emergency ICU admissions were identified by searching PICANet data for admissions with ADTYPE = 2 or ADTYPE = 4 (emergency admissions). Entering the dying phase was set to 28 days before the date of death, with these determined as set out in the next section. The most severe stage of condition was recorded for each person in each year. In the absence of other data (i.e. at the start of follow-up) children and young people were assumed stable.

Deaths and cause of death

Dates of death (if existing) were derived from four sources:

- ONS-derived mortality data (ADDE - date of death)
- The Congenital Anomaly Register (CARS - date of death)
- The inpatient data (PEDW - discharge date for admissions where discharge outcome was death)
- A&E data (EDDS - discharge date where discharge outcome was death)

Use of sources other than the ONS-derived mortality data (ADDE) enabled the inclusion of deaths that had not yet been formally registered, a process which can be delayed.(13) Where there were conflicts, but deaths were recorded in ADDE and other sources, ADDE data were preferred.

For those persons in Wales known to have died, they were also categorised as having or not having a life-limiting condition present (matching the ICD10 and/or Read code frameworks), considered present when:

- There was a life-limiting condition identified from inpatient (PEDW), primary care (WLGP) or congenital anomaly (CARS) data as described above.

OR

- There was *any* mention of a life-limiting condition in the death record (ADDE).

Previously derived demographic data were used to summarise deaths with and without a life-limiting condition by region and deprivation category. Age at death was assigned either from the death record or the age in the year of death if death was ascertained from other sources.