Report on DicData group of Projects

May 2012

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Preface

In our presentation to Children's Hospice UK in June 2010, we represented the 'DicData' group of projects as further steps in a research programme already started by a team from Bangor and Cardiff Universities. Funding from CHUK has enabled us to extend that collaboration to include one of the two Children's Hospices in Wales, Ty Gobaith, with the support and encouragement of the second, Ty Hafan,

The overall objective of the project was to bring together academic and clinical skills to generate, design and carry out a clinically relevant project, in order to address in a scientifically robust way research questions that are important both to users and to providers of palliative care services. In this way, we hoped to ensure that research remains firmly pegged to clinical needs of users and providers, so that its conclusions can usefully inform children's hospice and palliative care services, allowing them to remain sensitive to the needs of families, and facilitating their goal of equitable access. Alongside that, our aim was to begin to expand research infrastructure within Children's Hospices themselves, both through collaboration and by beginning to enhance research skills and interest among those working with patients.

The DicData series of projects has already achieved many of these aims. In Project One, we completed development of the Dictionary of Life-limiting Conditions in Childhood that had been the focus of our initial research collaboration. In Project Two, we used the Dictionary in conjunction with the Millennium Cohort Study (MCS) to estimate the maximum number of children in the UK population who might benefit from hospice and palliative care services. In Project Three, we focused on those families, drawing conclusions about the emotional and psychological impact of caring for a child with an LLC from secondary analysis of MCS data. This work was supplemented, thanks to an

additional research development grant, by Project Four, an integrative review of the literature by Sonia Ezergailis, now working at Ty Gobaith Children's Hospice.

We are continuing to a fifth project that will extend those aims further through a 'first in to research' project. Kate Barron is a children's nurse at Ty Gobaith Children's Hospice with an interest in developing her research skills. Her research involves mapping the psychological problems we have identified onto a set of previously conducted interviews with parents, children and professionals. It will form the fifth and final project in the DicData series. It is not yet complete, though we hope to complete the majority of it before the end of July 2012.

Like any project, we have encountered some unexpected challenges. There were unavoidable delays in agreeing contracts between University and Hospice in the early stages of the project. More recently, in analysing our data we became aware of a limitation in the Millennium Cohort Study data, which we now have to accommodate by modifying our analysis.

We are most grateful for Children's Hospice UK, now Together for Short Lives, without whom this important series of projects, and the opportunities it has given us to establish research links between clinical and academic institutions, could not have happened. We are confident that those good working relationships, the skills we have developed, and the potential they offer for ongoing collaboration between academic institutions and Children's Hospices, will continue far into the future.

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Project 1:

Life-limiting conditions among children in Wales: use of a 'Dictionary' towards defining service needs[1]

[1] The DicData team gratefully acknowledge the help of Dr Mary Devins, now Consultant in Paediatric Palliative Medicine in Dublin, Ireland, for her help in obtaining the death certificate data.

Executive summary of project 1

Our aim:
Our aim was to develop a tool for defining conditions that are life-limiting by ICD10 code, and use it to obtain data about childhood deaths in Wales in relation to palliative care.

What we did:
We compiled a list of diagnoses at referral to children hospices and to specialist palliative medicine services. Duplicates and diagnoses that were not LLC or were not primary were removed. Death Certificate data of all children in Wales between 2002 and 2007 were analysed to establish the proportion of deaths caused by LLC.

Headline findings:
- We described 376 different LLC. Around half deaths among children in Wales were from LLC (569/1052, 54%).
- Many LLC diagnoses obtained from death data did not appear in referrals to palliative care services, suggesting under-recognition of these disorders.
- The DLLCC is a practical tool for identifying LLC using ICD10 codes that facilitates extraction and analysis of data from existing sources in respect of LLC, offering the potential for rapid and precise studies in paediatric palliative care.

Comments:
An important incidental finding in this project was that a significant proportion of diagnoses, particularly from hospices, were not true LLC, often because there was confusion between the condition itself and symptoms of, or treatment for, the condition. This highlights an important potential for categorisation errors in using service data for epidemiological studies.

Status:
The manuscript was declined by Pediatrics but is currently being considered for publication in Archives of Disease in Childhood.

1.1 Introduction

In order to describe the population of children who need palliative care, the importance of defining palliative care in children has been highlighted.[1-8]

Palliative care is defined, not by organ system, but by the needs of an individual child and family, and any definition must be sufficiently flexible to accommodate this. The Royal College of Paediatrics and Child Health (RCPCH), working with the Association for Children's Palliative Care (ACT) in 1997, defined the concept of life-limiting condition[5] through a series of generic descriptions, but did not attempt to name specific diagnoses except as exemplars. If, however, children are to have the same access to specialist palliative interventions as adults currently enjoy, service developers must engage commissioners. That requires a precise understanding of the numbers of children who need services.

To establish numbers in Wales, we defined a list of several hundred of the most common specific conditions that were life-limiting according to the ACT/RCPCH definition. We then assigned ICD10 labels to each to create a 'Dictionary of Life-limiting Conditions in Childhood' (DLLCC), which allowed conclusions about children with LLC to be drawn from existing population data in Wales.

We consider the implications of these findings, and some of the wider applications of the Dictionary in taking forward service and research developments in children's palliative care.

1.2 Materials and methods

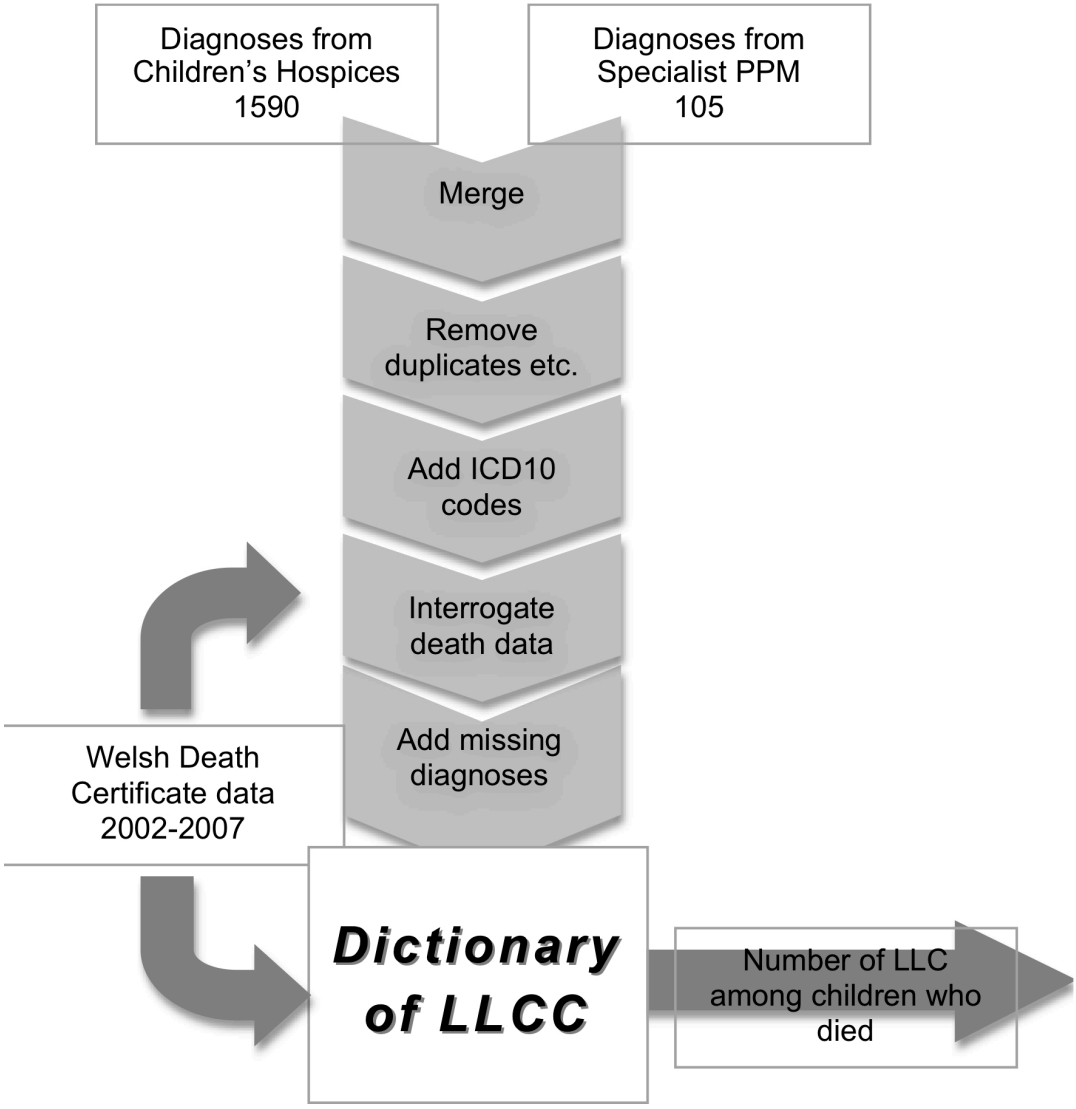

Figure 1.1: Schematic showing stages in the study. A preliminary list of life-limiting conditions was drawn up by combining diagnoses among patients accepted by children's hospices, with those accepted by specialist paediatric palliative medicine services. ICD10 codes were assigned to the diagnoses. Comparison of the merged list with death certificate data allowed refinement of the list, resulting in the first edition of the 'Dictionary of LLCC' which was then used to identify life-limiting conditions among all causes of death in a five-year period in Wales.

Diagnostic labels among children with life-limiting conditions were collected from two sources (Figure 1.1). First, diagnoses of children admitted to children's hospices were provided by Children's Hospices Hospice UK (CHUK), the

membership organisation for Children's Hospices in England and Wales. The diagnoses emanated from the five children's hospices that were using a standardised data collection tool developed by Chase Hospice (Esplen, personal communication 2010). Second, diagnoses of children accepted by specialist paediatric palliative medicine services were obtained from tertiary service based at the Children's Hospital in Cardiff. Neither source used any formal disease classification system.

The two lists of diagnoses were merged. The combined list was edited, removing duplicate diagnoses and descriptions that were not truly diagnostic labels. Duplicates occurred either when two diagnoses were exactly the same, or when two or more terms were used to describe the same condition (e.g., trisomy 13 and Patau's syndrome). Non-diagnoses included terms that had led to referral, but were deemed by the investigators to be modes of death rather than diagnosis (e.g., apnoea), were incidental to the life-limiting diagnosis (e.g., anaemia), or were treatments for the life limiting diagnosis (e.g., tracheostomy). Finally, diagnoses were removed if they could not be described by one or more of the ACT/RCPCH categories and therefore, by the definition used for this study, were not life-limiting conditions.

A diagnostic label and code from the International Classification of Disease (ICD10) was assigned by the investigators to each diagnosis on the list (apps.who.int/classifications/apps/icd/icd10online/).

The combined list of LLCs built from evidence in practice was then compared with aggregated anonymous death certificate data for all deaths in Wales between 0 and 19 years between 2002 and 2007, obtained from Public Health Wales Observatory.[9] The number of child deaths in Wales caused by LLC (according to the study definition) over that five-year period was established. LLC that did not already appear in the DLLCC, were added to it.

1.3 Results

1590 diagnoses from children's hospices and 105 from specialist palliative medicine were combined. 1319 diagnoses were removed because they were duplicates, not palliative, or not diagnoses.

There were 1052 deaths in childhood between 2002 and 2007 (tables 1.1-1.3).

Number of deaths	Diagnosis	ICD10 code
26	Cerebral palsy, unspecified	G80.9
21	Meningococcaemia, unspecified	A39.4
15	Acute myeloid leukaemia	C92.0
13	Malignant neoplasm: Brainstem	C71.7
11	Septicaemia, unspecified	A41.9
11	Acute lymphoblastic leukaemia	C91.0
11	Epilepsy, unspecified	G40.9
9	Malignant neoplasm: Brain, unspecified	C71.9
9	Muscular dystrophy	G71.0
8	Malignant neoplasm: Connective and soft tissue, unspecified	C49.9

Table 1.1: Top ten causes of death that are describable by one of more of the ACT/RCPCH categories and therefore considered 'life-limiting' conditions.

Number of deaths	Diagnosis	ICD10 code
60	Birth asphyxia, unspecified	P21.9
19	Hypoplasia and dysplasia of lung	Q33.6
16	Necrotizing enterocolitis of fetus and newborn	P77
16	Congenital diaphragmatic hernia	Q79.0
10	Edwards' syndrome, unspecified	Q91.3
8	Congenital malformation of heart, unspecified	Q24.9
7	Persistent fetal circulation	P29.3
4	Congenital renal failure	P96.0
4	Hypoplastic left heart syndrome	Q23.4
3	Severe birth asphyxia	P21.0
3	Patau's syndrome, unspecified	Q91.7

Table 1.2: Top eleven diagnoses (those accounting for three or more deaths) causing death among neonates describable by one of more of the ACT/RCPCH categories and therefore considered 'life-limiting' conditions. In total, there were 169 deaths from 24 diagnoses.

ICD10 chapter heading	Description	Number deaths (%)
C or D	Neoplasms, diseases of the blood and blood-forming organs and certain disorders involving the immune mechanism	104 (25%)
A	infectious and parasitic diseases	50 (12%)
E	Endocrine, nutritional and metabolic disease	24 (6%)
G	Diseases of the nervous system	88 (21%)
I	Diseases of the circulatory system	36 (9%)
J or P	Diseases of the respiratory system, certain conditions originating in the perinatal period	30 (7%)
Q	Congenital malformations, deformations and chromosomal abnormalities	54 (13%)

Table 1.3: Contribution of different categories of life-limiting condition to death in childhood (outside neonatal period). The range of diagnoses is much wider than in adult specialty, but cancer is still the commonest single group.

Of these, 569 (54%) were deemed to be due to an LLC. Of 382 causes of death on certificates, 186 (49%) were not considered true life-limiting conditions as they could not be described by any ACT/RCPCH category.

Only seven LLC diagnoses caused ten or more deaths (table 1.1). Among deaths from LLC, the ten commonest diagnoses accounted for 32%, while the 136

diagnoses that caused one or two deaths accounted for 25%. Table 1.3 shows causes of death by ICD10 chapter. The majority occurred from a small number of life-limiting conditions. Malignancy (25%) and neurological conditions (21%) were the most frequent (table 1.3).

Among neonates (table 1.2), there were 169 deaths from life-limiting conditions, of which 92 (54%) were from ICD10 chapter P (certain conditions originating in the perinatal period), and 73 (43%) were from chapter Q (congenital malformations, deformations and chromosomal abnormalities). The total number of deaths from congenital malformations, deformations and chromosomal abnormalities was therefore 127, or 22% of total deaths in both age groups. 97% deaths in the neonatal period were accounted for by conditions in only two ICD10 chapters.

1.4 Discussion

The background to this study was the difficulty defining palliative care in children and thus providing data suitable to contribute to planning for services. In children, the term 'life-limiting condition' encompasses non-malignant as well as malignant conditions and the range of conditions is wide. In the UK and, increasingly, elsewhere, LLC in children are conventionally classified by the ACT/RCPCH system,[2, 4, 6, 8] which relies for its validity on assumed commonality among the courses of diseases within each of four categories. There is currently little evidence to support this.[10] Moreover, where registration criteria need to be precisely defined, the ACT/RCPCH categories as they stand are too vague and need to be supplemented by identifying precise diagnoses.

We are not, of course, the first to recognise the need for specific data in service development. Lists of life-limiting conditions have been compiled before, notably by Knapp (personal communication 2011), Craig[8] and Feudtner.[11,12] In order to support service development in Great Britain, we have tried to develop

the concept by expanding the number of diagnoses and drawing on a range of UK sources. Most importantly, we tried to ensure the data could be used by service commissioners by using a well-recognised and widely accepted 'yardstick' for deciding whether a given condition is life-limiting. The virtue of the ACT/RCPCH system is that it captures the diversity of conditions that can limit life; our aim was to obtain useful precise data without losing that virtue.

The range of LLC causing death in the neonatal period was startlingly narrow (table 1.2). Only five conditions accounted for almost three-quarters of these deaths (121 out of 169, 72%). A detailed care pathway that provides a standardised approach to palliative care for neonates, analogous to the Liverpool Care Pathway in adults,[13] could be appropriate and effective.[14] This approach has been controversial in adult services[15] and, outside the neonatal period, the sheer diversity of LLC (table 1.1) probably makes it impractical for children.

Diagnoses that emanated from hospices were not the same as those from specialist PPM services. Children's hospices typically offer short respite stays and are often nurse-led. In contrast, specialist PPM services are based around availability of specialist medical services. Although the two populations clearly significantly overlap, they are not precisely co-terminous,[16] and combining them therefore further expanded the number of diagnoses on the list.

Most individual LLC caused death only once in the study period, and few diagnoses (5 in neonates, 7 in older children) caused it 10 times or more (tables 1.1 and 1.2). At the same time, nearly one third of deaths were accounted for by only ten different LLC, confirming clinicians' impression that, while the range of possible LLC is wide, it is possible to identify a relatively small number of diagnoses whose symptom management should form the core of a specialist palliative care skillset.

Of 420 deaths from LLC outside the neonatal period, 75% were from conditions other than cancer. This is higher than in studies that have relied on reporting by paediatricians,[16] suggesting there has been under-recognition of the life-limiting nature of non-malignant conditions.

In this study, a condition was life-limiting if it could be described by one or more of the ACT/RCPCH categories. On the face of it, it seems remarkable that around half of the conditions from which children died were not so described. One explanation may be the unknown validity of the records of causes of death, and replication of these findings using other data sources would be valuable. A second and intriguing explanation, however, may be that the ACT/RCPCH categories are currently too restrictive. The categories were developed on the basis of expert opinion, and have in common a high probability that a child will not survive into adulthood. This is sometimes expressed by the 'surprise' question: 'Would you be surprised if this child were to survive into adulthood?'. The ACT/RCPCH categories identify those who need access to PPC services (including financial and psychological support, symptom control and support after death), though at any given time only a proportion will have 'active' needs.[5]

It could be argued that children with many diagnoses that are excluded from the Dictionary require care that is, in effect, palliative. The commonest of these outside the neonatal period (table 1.3) is traffic injury.[17] Although accidents do not fit an ACT/RCPCH category, PPM services could have a valuable role (e.g. with difficult decision-making in intensive care). Perhaps this indicates a potential value in extending the ACT/RCPCH categories to reflect the broader role that might be played by PPM services.

Conditions such as diabetes and epilepsy are usually incurable and require the same approach and 'ethos' as palliative care. There is certainly a risk of drawing an arbitrary distinction between palliative care and what is simply good clinical practice in children. Nevertheless, those working in the field recognise a

population of children within this wider group who are at high risk of death during childhood, and in whom complex symptom control is a frequent clinical challenge. It is that population that the Dictionary aims to help to identify.

There are inevitable limitations to a study of this nature. Although conditions were only considered life-limiting if they could be described by one or more ACT/RCPCH category, the categories themselves were not designed to be exhaustive, and there is an unavoidable degree of subjectivity on the part of investigators in making that judgement.

In the current study, diagnoses were added to the DLLCC if they caused death in the study period and, despite fulfilling the study definition of a LLC, did not already appear in it. It might appear that the DLLCC is therefore dependent on death certificate data. In fact, however, since diagnoses were not added unless they fulfilled the study criterion for an LLC, this dependence is no greater than the dependence of both on that criterion. No diagnoses were included unless they were life-limiting according to the study definition, so that the effect of adding more diagnoses to the DLLCC can only have been to improve approximation to the actual sum total of all LLC.

It is not possible to list every possible LLC in a Dictionary: there will need to be a mechanism for adding new diagnoses as they become apparent. Publically available data recorded on death certificates is limited to the principal causes of death. It is possible that small numbers of children with LLC who died from incidental causes were not identified in this study. Removing duplicate diagnoses is complex, since several clinical labels can attach to a single diagnosis. Furthermore, conditions such as cerebral palsy may limit life-span in some children but not others.

There are important disease classifications other than ICD10, including Read Coding, used in primary care,[18] and the ACT/RCPCH categories

themselves.[5] Assigning ACT/RCPCH and Read Categories to ICD10 diagnoses could be a further stage in development of the Dictionary of LLCC.

This study demonstrates the number of children whose cause of death was a LLC, rather than the much larger number living with them (prevalence) or even those with a LLC who die of another cause. More complete prevalence data are necessary to underpin rational development of clinical services. Their long natural history means that to be able to show prevalence of LLC reliably, studies would ideally require a prospective population study, in which children were identified across the whole sample. Given the low prevalence of LLCs in a typical population, this will in practice require analysis of existing population-based databases, such as the Millennium Cohort Study. By listing LLC by ICD10 code, the DLLCC facilitates such analysis.

The Dictionary has a number of immediate practical applications where the sub-population of children with LLC needs to be identified within larger groups such as those with complex chronic disability or other chronic illness. It can rationally underpin fair admission and referral criteria for children's hospice services, and help evaluate the magnitude of the need for specialist palliative medicine and palliative care services for children by institutions within the National Health Service. In countries such as the USA with a private healthcare system, the Dictionary can inform funding decisions among insurance companies. It can also facilitate robust governance and record-keeping by those providing palliative care, by allowing a definition of palliative care derived from a standard that has been largely agreed.

The Dictionary has potentially important applications for research in paediatric palliative care. As the number of epidemiological studies increases, the need to compare their findings becomes both more important and more difficult without a standardised definition of LLC. To define the population of children needing palliative care in an essential first step in considering any research question that

impacts specially on that group. The Dictionary has already been used for this purpose in research[19] and service development.[20]

Finally, the Dictionary can potentially allow critical evaluation of the ACT/RCPCH categories themselves, allowing research that can inform amendments and improvements to what are seen, in the UK and increasingly beyond, as the standard definition of what constitutes a 'palliative' diagnosis.

1.5 Conclusions

The authors have compiled a list of ICD10 diagnoses that have been judged by professionals working in paediatric palliative care to be life-limiting. We drew on admissions to children's hospices on the one hand, and referrals to specialist paediatric palliative medicine on the other. As a final step, death certificate data were used to revise and then to apply the list for a practical purpose.

Around half of all childhood deaths in the study period were from LLC. Three-quarters of LLC were non-malignant. Outside the neonatal period, the range of conditions was wide, with few conditions accounting for more than 10 deaths in the five-year study period. In neonates, by contrast, the range of LLC was narrow, with most LLC being in only two ICD10 chapters.

The DLLCC can never be exhaustive. As new diagnoses become apparent, expansion will be important and should be the basis for further studies, which should also attach Read and ACT/RCPCH categories.

By defining a list of ICD10 codes that fulfil ACT/RCPCH criteria, for the first time the DLLCC allows analysis of existing clinical databases, paving the way for rapid establishment of prevalence data that would otherwise have been impractically slow. While it will continue to need refinement, the DLLCC is a key tool for rational service development in children's palliative care.

1.6 References

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Project 2:

Prevalence of life-limiting conditions in children aged 3, 5 and 7 years: secondary data analysis of the Millennium Cohort Study

Draft 28May12

Executive summary of project 2

Our aim:
Our aim was to establish true prevalence of LLC in early childhood and describe examine sociodemographic correlates of LLCs.

What we did:
We used the Dictionary to identify all children with LLC in the Millennium Cohort Study (MCS) at three ages (three, five and seven years), extracted data from the MCS in respect of relevant demographic characteristics such as relationship, employment and socio-economic status, and compared those data between families with LLC and those without.

Headline findings:
- At all three ages, prevalence in our study was much higher than in any other study at approximately 2% (or 200/10000 children).
- Families caring for children with LLC are more likely to be single, poor and unemployed than those without.

Comments:
It is likely that three reasons contributed to the high prevalence in our study:

- We identified children who were not already accessing services, as well as those who were.
- By studying a narrow range of ages, we have uncovered indirect evidence that prevalence fluctuates across the age groups in childhood.
- A limitation in the MCS in relation to the ICD10 chapters means that our study overestimates prevalence.

Unfortunately, it is impossible to know the relative contributions made to our high estimate of prevalence by each of these three factors, which limits the interpretation we can put on the findings as they stand. Correcting the data to minimise the impact of the chapter limitation issue in the MCS will make that clearer.

Status:
This project analysis is complete and a manuscript prepared, but we have had to delay submission since we will need to revise the final analysis to address some of the limitations associated with being able to use only ICD Chapter level categories within the MCS. Our prevalence estimates will still be much higher, and we believe this observation will be a significant contribution to the debate about LLC in children.

Draft 28May12

2.1 Introduction

Establishing the prevalence of life-limiting conditions in children is both important and challenging. Its importance lies in the need to demonstrate to commissioners that development of palliative care services is necessary. Its challenge lies in the difficulty identifying all children who would potentially benefit from the availability of such services. Many of the key terms are difficult to define, and it is problematic in practice to identify all the children who should be recruited to any study.

In this project, we addressed the first of those difficulties by utilising the dictionary of LLC conditions (DLLCC) we had developed in project 1. This enabled us to define with some clarity the population of children whose problems we were trying to establish. The second difficulty can only be overcome by utilising a cohort study design, which is designed to ensure that the population under study is not different from the general population in any way that is significant to the outcome of the study. We performed secondary analysis of data gathered on a cohort of children recruited into the Millennium Cohort Study because of the date of their birth.

Since date of birth has no plausible impact on the likelihood of any individual child developing a LLC, this design allows conclusions to be confidently drawn in respect of prevalence in the population at large. Studies of prevalence that have to rely on identifying patients already known to services provide good estimates of the *minimum* demand for services, because the true prevalence must be that or greater. To establish the *maximum* number of children who could potentially be affected by a LLC and thus benefit from children's care services, however, a cohort study design is essential.

Draft 28May12

2.2 Materials and methods

This is a secondary analysis of data available in the second, third and fourth surveys (ages 3, 5 and 7) of the Millennium Cohort Study. The MCS was designed to identify a population-representative sample of children in the UK, born in the new millennium and to follow them up prospectively. Participants were identified through the Child Benefit Register. The child benefit is a non-means tested benefit available to all UK children. Participant identification was geographically clustered to include all four countries (England, Wales, Scotland, Northern Ireland), and disproportionately stratified to over-include ethnic minority and disadvantaged children (1, 2). Over-sampling was done to ensure adequate representation of these groups in the survey (1). Children and families were selected from 398 randomly selected electoral wards in the UK.

At age 3, there were 15,590 children who participated in MCS2; at age 5, 15,246 children were included (MCS3), and at age 7, 13,857 children were followed up (MCS4). All analyses conducted and reported below used appropriate weights to account for the disproportionate stratification of ethnic minority and disadvantaged children along with attrition/non-response.

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2.3 Results

Definition of terms used in this study

Point Prevalence: Number of children with LLC at a specific time point as a proportion of the total number of population at that time point

Persistence: Number of children with an LLC at both time points as a proportion of number of children with LLC at the first time point

New cases: Children with an LLC at T2 who did not have an LLC at T1 as a proportion of the number of children with available data at both time points (total population for this ratio)

Incidence: Number of new cases, expressed as a proportion of number of children with available data at both time points MINUS number of children who had LLC at start point ('population at risk'). Also known as Cumulative incidence, or incidence proportion (3).

Prevalence

Prevalence of LLC in the three age groups is shown in table 2.1. At age 3, this prevalence corresponds to a weighted count of approximately 321 children, of whom 305 have one LLC, 14 have two LLCs and 2 have three LLCs. At age 5, the weighted estimate is 318 children of whom approximately 20 have two LLCs (6.22%) and 298 have one LLC (93.78%). At age 7, the weighted count is 304, of whom 290 have one LLC and 14 have two LLCs.

There was no observable change in prevalence over time. McNemar's paired groups chi-square tests showed no significant difference between prevalence at ages 3 and 5 (χ^2= .03, p=.911), 5 and 7 (χ^2= 1.27, p=.285), or 3 and 7 (χ^2= 2.35, p=.143)

	Unweighted prevalence	**Weighted prevalence**	**Unweighted base population**
Age 3	1.86% (1.65% to 2.07%)	2.04% (1.80% to 2.29%)	15,590
Age 5	1.92% (1.70% to 2.13%)	2.04% (1.78% to 2.30%)	15,246
Age 7	2.00% (1.77% to 2.23%)	2.19% (1.87% to 2.51%)	13,857

Table 2.1: Prevalence at ages 3, 5 and 7 years.

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Frequency of LLCs by cohort

The ten most frequent LLCs within each time point (ages 3, 5, and 7) were identified. These are presented in tables 2.2a-2.2c, accompanied by the corresponding ICD-10 name.

Age 3	**ICD-10 name***	**Weighted frequencies**
1 Q21	Congenital malformations of cardiac septa	**12.64%**
2 G40	Epilepsy	**9.06%**
3 F84	Pervasive Developmental Disorders	**7.61%**
4 G80	Cerebral Palsy	**7.41%**
5 J98	Other respiratory disorders	**7.01%**
6 Q60	Renal agenesis and other reduction defects of kidney	**5.02%**
7 Q74	Other congenital malformations of limbs	**4.98%**
8 R06	Abnormalities of breathing	**4.70%**
9 Q82	Other congenital malformations of skin	**3.96%**
10 Q87	Other specified congenital malformation syndromes affecting multiple systems	**3.04%**

Table 2.2a: The 10 most frequent LLCs at age 3 years (*Description based on current version ICD-10 2010)

Age 5	**ICD-10 name***	**weighted frequency**
F84	Pervasive Developmental Disorders	**30.40%**
G40	Epilepsy	**16.76%**
J98	Other respiratory disorders	**8.61%**
G80	Cerebral Palsy	**7.04%**
R06	Abnormalities of breathing	**5.81%**
Q21	Congenital malformations of cardiac septa	**5.01%**
Q87	Other specified congenital malformations syndromes affecting multiple systems	**3.60%**

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Q82	Other congenital malformations of skin	**2.48%**
Q79	Congenital malformations of the musculoskeletal system not elsewhere classified	**1.86%**
Q85	Phakomatoses, not elsewhere classified	**1.54%**

Table 2.2b: The 10 most frequent LLCs at age 5 years (*Description based on current version ICD-10 2010)

Age 7	ICD-10 name*	Weighted frequency
F84	Pervasive Developmental Disorders	37.41%
G40	Epilepsy	18.30%
G80	Cerebral Palsy	5.75
Q21	Congenital malformations of cardiac septa	3.77
N25	Disorders resulting from impaired renal tubular function	3.64
F80	Specific developmental disorders of speech and language	3.02
Q82	Other congenital malformations of skin	2.94
J98	Other respiratory disorders	2.43
Q87	Other specified congenital malformations syndromes affecting multiple systems	2.19
Q85	Phakomatoses, not elsewhere classified	1.60

Table 2.2c: The 10 most frequent LLCs at age 7 years (*Description based on current version ICD-10 2010)

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Demographic characteristics of children with a LLC

Variable	LLC	NonLLC	Difference
Child characteristics			
Child male	55%	51%	F=1.69, p=0.193
Child ethnic status:			
White	87%	87%	
Mixed	3%	3%	
Indian	2%	1%	
Pakistani/Bangladeshi	5%	4%	
Black or Black British	2%	3%	
Other (inc. Chinese)	1%	1%	F=0.179, p=0.959
Family Characteristics			
2+ siblings	30%	28%	F=0.19, p=0.661
Large family (5+ people in household)	29%	30%	F= .014, =0.906
Single-parent household	25%	18%	F=8.41, p=0.004
If 2- parents, % married	79%	78%	F=0.487, p=0.609

Whether main respondent is mother	99%	99%	
Maternal age	31 (.44)	32 (.12)	F=5.20, p=0.02
Social and Economic indicators			
Subjective poverty (finding it quite/very hard to manage)	12%	10%	1.13, p=0.282
Area Deprivation (lowest 20% of IMD)	23%	23%	.0168, p=0.897
Both parents unemployed	10%	7%	4.798, p=0.029
Income Poverty (<60% median UK equivilised income)	35%	29%	3.93, p=0.048
High deprivation (2+ adverse SEP indicators)	19%	17%	F=0.36, p=0.549

Table 2.3a: Demographics of families of children with (LLC) and without (NonLLC) at age 3.

	LLC	**NON-LLC**	**Difference**
Child characteristics			
Gender Male	63%	51%	sig
Child ethnic status: White Mixed Indian Pakistani/Bangladeshi Black or Black British Other (inc. Chinese)	 86% 3% .5% 4% 3% 2%	 87% 6% 2% 3% 3% 1%	 1.96, p=0.111
SEP			
Subjective poverty (finding it quite/very hard to manage)	7%	11%	2.09, p=0.149
High area Deprivation (lowest 20% on IMD)	22%	22%	non-sig
Neither parent working	33%	20%	highly sig
Income Poverty (OECD)	31%	31%	non-sig
High Deprivation (2+ adverse SEP indices)	28%	24%	1.77, p=0.185
(Cumulative deprivation since 9 months)	2.31 (.18)	1.99 (.07)	F=3.94, p=0.05
Family Characteristics			
2+ siblings	35.92%	35.17%	non-sig
Large Family (5+ members in HHD)	34.73%	32.47%	non-sig
Single Parent household	29%	20%	10.10, p=0.002
When 2 parents, are they married?	75.54%	76.22%	non-sig
Mum main respondent	98%	98%	
Maternal age	34 (.41)	34 (.12)	F=0.48, p=0.487

Table 2.3b: Demographics of families of children with (LLC) and without (NonLLC) at age 5.

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Variable	LLC	NonLLC	Difference
Child characteristics			
Child male	65%	51%	17.47, p<.001
Child ethnic status: White Mixed Indian Pakistani/Bangladeshi Black or Black British Other (inc. Chinese)	 88% 5% 0.1% 4% 3% 0.4%	 86% 3% 2% 5% 3% 1%	 2.24, p=0.06
Family Characteristics			
2+ siblings	46%	40%	2.97, p=0.08
Large family (5+ people in household)	36%	39%	.55, p=0.458
Single-parent household	33%	23%	11.38, p=0.001
If 2- parents, % married	68%	72%	5.83, p=0.004
Whether main respondent is mother	98%	97%	.25, p=0.614
Maternal age	35 (.39)	36 (.12)	2.76, P=0.097
Social and Economic indicators			
Subjective poverty (finding it quite/very hard to manage)	16%	13%	1.63, p=0.202
Area Deprivation (lowest 20% of IMD)	25%	22%	.78, p=0. 377
Both parents unemployed	10%	7%	2.55, p=0.110
Income Poverty (<60% median UK equivilised income)	36%	29%	4.34, p=0.038
High deprivation (2+ adverse SEP indicators)	24%	20%	2.02, p=0.156

Table 2.3c: Demographics of families of children with (LLC) and without (NonLLC) at age 7.

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Incidence, Persistence and New Cases

These are shown in table 2.4. There was significantly less persistence between three and five years than between five and seven years (in the 5-7 period (χ^2= 19.66, p<0.001, McNemar's chi-square). There was, however, no significant difference in the rate of new cases emerging in these two time periods (χ^2=2.11, p=0.165).

	3 to 5 years (95% CIs)	5-7 years (95% CIs)	3-7 years (95% CIs)
% Persistence	43.31% (35.78% to 50.84%)	61.55% (52.62 to 70.48%).	40.91% (33.07% to 48.75%
% New cases	1.11% (.89% to 1.33%)	.93% (.74% to 1.12%)	1.32% (1.07% to 1.58%)
Incidence	1.13% (.91% to 1.36%)	.95% (.91% to 1.36%)	1.35% (1.09% to 1.61%)

Table 2.4: Incidence, persistence and percentage of new cases (with 95% Confidence Intervals) in each of the three age cohorts.

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2.4 Discussion

This is the first study to use secondary analysis of cohort data to examine LLC in childhood. Previous studies have had to rely on reporting (4-6), meta-analysis (7, 8), service usage data (9, 10) or a combination of these (11, 12). By analysing the MCS, we were able to study the same population at ages 3, 5 and 7 years.

The population did not change significantly over that period, although there were some minor differences caused mainly by changing numbers of children available for data, which fell from 15,590 at age 3, to 15,246 at age 5 and finally to 13,857 at age 7. An important observation from our study is that 6.22% (more than one child in twenty) who has one life limiting condition may also have a second. This emphasises the potential complexity for parents and for health professionals in caring for children with LLC.

The most important conclusion of this study was that the prevalence of life limiting conditions (LLC) in the population at all three times it was studied was approximately 2%, or 200 per 10,000. That is higher than in other studies, which have converged on a figure of between 10 and 40 per 10,000 (4-12).

It seems likely this has three explanations. Prevalence from a cohort study represents the number of children in the population who have a life limiting condition. This is logically greater than the number of children with a LLC who actually access palliative care services. It needs to be distinguished carefully, however, from the number of children who will need input from palliative care services at any one time. Given the definition that we used, all the children identified by our study are likely to need palliative care at some time in the course of their condition. It does not follow, however, that they all need it all the time; in fact that is highly unlikely. The figure we have represented here, therefore, is the maximum number of children who could logically need access palliative care services. The number who will need to do so at any one time is likely to be a

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fraction of this.

This high evidence might also provide evidence of a change in prevalence over the course of childhood. It is intuitively implausible that the prevalence of LLC is constant over the whole range of age groups usually considered in prevalence studies (typically 0-19 years). The diagnosis of an LLC is usually made in the neonatal period, or in the first few months and years of life as delayed developmental milestones become apparent. It would be relatively unusual for a LLC to be diagnosed for the first time in late childhood or adolescence, though cancer would be an exception. The number of children living with LLC is also likely to change over the course of childhood. Conditions such as genetic anomalies that are diagnosed in the neonatal period often lead to death within weeks or months, while those with a natural trajectory of many years, such as cerebral palsy, are often diagnosed in infancy and those such as cancer that can be diagnosed at any point in childhood will typically persist for some years.

The result of these variations in the rate of new diagnosis on the one hand, and of death on the other, is likely to be a complex pattern of fluctuating prevalence over the age groups of childhood that has not previously been identified. By studying a narrow range of ages in the context of a cohort study, our study may provide the first evidence for it. We have shown greater fluidity among diagnoses between ages of 3 and 5 than between 5 and 7 (table 2.4).

A further factor contributing to that high prevalence figure, however, relates to a limitation of the Millennium Cohort Study in itself, which limits our interpretation of these data and. The Dictionary, like the ICD10 classification itself, offers precise descriptions of individual conditions under chapter subheadings (ie. up to 4-digit ICD-10 codes), grouped together under more general chapter headings. Unfortunately, the MCS does not do this; it records only chapter headings (ie. only 2-digit codes). Within the same chapter, therefore, it is likely that some conditions that are not life-limiting will be grouped for analysis as though they were. Inevitably, this means that secondary analysis of the MCS data will result

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in overestimation of prevalence.

The most obvious example of this in our study is in considering the chapter F84 (Pervasive Developmental Disorders). Conditions in chapter F84 frequently occurred among LLC (table 2.2a). According to the Dictionary, however, only one diagnosis in chapter F84 is actually an LLC; that is, Rett Syndrome, F84.2. Chapter F84 includes only one genuine life limiting condition in a chapter of diagnoses that affect a large number of children. To minimise the effect of this limitation, the solution would be to remove all F84 diagnoses from our analysis. Removal of the F84 code is warranted because of the relatively high frequency with which it appears in the population and because removing Rett's Syndrome in this way from the list of LLC fortunately does not impact significantly on the conclusions we can draw, since it is rare and in any case it is not agreed among clinicians that Rett's should be considered a LLC. This is likely to result in a decrease in LLC prevalence rates. Nevertheless, until the MCS, and other national cohort studies, begin to record data relating to ICD10 subheading as well as heading, it is inevitable that any secondary analysis such as ours will overestimate the prevalence of LLC.

There was no significant increase in point prevalence over the course of the three analyses that were made (tables 2.1 and 2.2a-c). Although there appears to be a trend towards an increase in the age 7 group (albeit non-significant, table 2.1), this is again likely to be an artifact of the MCS in relation to F84 diagnoses. There is, however, a significant change in the distribution of LLC conditions between three and seven years, most marked between the ages of three and five, when congenital malformations of the heart and bowel give way to pervasive developmental disorders (tables 2.3a-c). This explanation is supported by the fact that an LLC diagnosed at age five is more likely still to be present when the child is seven than if the diagnosis is made at age 3 (table 2.4). It seems likely that the causes of this fluidity are complex, with some children receiving

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diagnoses late, and others dying from LLC shortly after a diagnosis made in the first few months or years of life.

The second set of conclusions we were able to draw from our data relates to characteristics of families caring to children with life limiting conditions, compared with those who did not. The only significant demographic difference between the two groups was that there were significantly more boys among the families of children with LLC. This is probably explained due to the influence of X-linked genetic conditions that can limit life, particularly Duchenne Muscular Dystrophy.

There were, however, significantly more single-parent families and/or families with both parents unemployed among those with life-limiting conditions. Children with LLC were more likely than those without to be cared for in a single-parent household at all three ages. It is not clear whether that is a result of relationship breakdown, or to strategies of living separately in order to optimise state benefits. At ages three and five, among families with LLC it was more likely that neither parent was employed, and income poverty was more likely than at ages three and seven. At ages five and seven a child with life limiting condition was more likely to be male than a child without, and at age three a child with a LLC was likely to have a mother who was significantly older than one without. Some of these differences are intriguing but probably have little impact on the lives of children and families, but the overall pattern that emerges from these data is that families coping with the care of a child with a LLC are more likely to be doing so single-handed, and to be struggling with financial difficulty, than those without.

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2.5 Conclusions

By using a cohort study design, we have been able to estimate prevalence of LLC in the population and to compare this when the cohort was three, five and seven years old. The prevalence of LLC in our study was much higher than previous estimates, even a study that incorporated the same Dictionary

alongside other methods to define patients for inclusion. Reasons for this include the capacity of our study to identify patients who were not known to clinical services, and the fact that we studied a narrow range of age groups, in which prevalence may be higher than in other age groups in childhood. Our study provides evidence that the distribution of life-limiting diagnoses affecting children changes more rapidly in early age groups than later. This supports the suggestion that prevalence of LLC may vary across the age-groups of childhood, perhaps because rate and timing both of diagnosis and of death evolve in complex ways over early months and years.

A limitation is that the MCS overestimates LLC in the population, because it records only ICD10 chapter headings, meaning that within specific chapters including a variety of disorders it cannot distinguish between LLC and non-LLC. Repeating our analyses, excluding chapter F84 will improve the overall validity of our findings, but we cannot entirely overcome the limitation inherent in the MCS data.

There are significant differences between characteristics of families with and without children with LLC that are relevant to the practical difficulties of caring for them. These include a greater likelihood of unemployment, poverty and being single parents.

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Project 3:

Wellbeing in families with a child with a life-limiting condition: Evidence from secondary analysis of a British Cohort

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Executive summary of project 3

Our aim:
To examine developmental outcomes in children with LLC (cognitive, behavioural and physical development), along with maternal well-being, and their potential association. the impact of behaviour and cognitive ability in children with a life-limiting condition on the well-being of their families.

What we did:
We analysed all data collected in the MCS3 that were relevant to the child's behaviour and the family's well-being (for example, life satisfaction), and compared those with and those without life-limiting conditions.

Headline findings:
- Behaviour problems, cognitive impairment and obesity were all more likely among children with LLC.
- Although general health status was less good, and psychological distress was higher, families withmothers of children with LLC did NOT report any less satisfaction with their lives than those without.
- Neither was caring for a child with a life-limiting condition associated with increase likelihood of serious psychological illness
- Behavioural problems, cognitive impairment and obesity were all more likely among children with LLC.

Comments:
This is important because it questions a model of universal vulnerability and victim-hood among families caring for children with LLC, while at the same time affirming the multiple sources of stress they are faced with.

Status:
This project is complete and a manuscript drafted. Since it was a more intuitive ordering, we elected to delay publication until after that of project 2. These results, like those of project 2, will need re-interpreting once we have addressed removed the F84 chapter limitation issue in the MCS data from our analyses.

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3.1 Introduction

The objective of caring for patients is ultimately to improve their well-being. The concept of well-being itself, however, is difficult to define, still more to measure. It is a translation of Aristotle's term *eudaemonia* that is both more precise and more complex than the traditional translation 'happiness'. Well-being more faithfully represents Aristotle's concept that *eudaemonia* describes an individual who is fulfilling the purpose for which they were created. It certainly encompasses an individual's capacity to be happy, but also other relevant capacities such as to relate to others, to be able to influence the environment around them, and to have a sense of *telos* - that is, a direction that gives purpose to life experience (Ryff 1989).

With this complex understanding, it becomes clear that well-being can be enhanced by events and circumstances that cause unhappiness, and even by unhappiness itself. In this project, we examine the impact on well-being of caring for children with life limiting condition. Its purpose was to examine any association with the behaviour problems, cognitive and physical development of children with life limiting conditions with the psychological and physical well-being of their mothers. To do this, we performed a secondary analysis of cohort data from the MCS, comparing data from families of a group of children with LLC and the remainder who did not.

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3.2 Materials and methods

This is a secondary analysis of data available in the third survey (age 5) of the Millennium Cohort Study (MCS3). The MCS was designed to identify a population-representative sample of children in the UK, born in the new millennium and to follow them up prospectively. Participants were identified

through the Child Benefit Register. The child benefit is a non-means tested benefit available to all UK children. Participant identification was geographically clustered to include all four countries (England, Wales, Scotland, Northern Ireland), and disproportionately stratified to over-include ethnic minority and disadvantaged children (Plewis 2007; Hansen 2010) Over-sampling was done to ensure adequate representation of these groups in the survey (Plewis 2007) . Children and families were selected from 398 randomly selected electoral wards in the UK. In MCS3, 19,422 participants were eligible, of which 15,246 actually participated (79.2% participation rate).

Participants

Among the 15,246 singleton children in MCS3, we identified 292 with a LLC (unweighted prevalence 1.89%, 95% CIs: 1.70% to 2.13%; Hain et al, Project 2). Weighting to account for the stratified clustering of the MCS design and nonresponse/attrition suggested a weighted prevalence of 2.04% (95% CIs: 1.78% to 2.30%). The remaining 14,954 children without a LLC were used as a comparison group. There were significantly more male children in the LLC group (63.3%) in relation to the comparison group (50.83%; χ^2:14.35, p=0.002), while as expected there was no variation in age. Hain and colleagues (Project 2) explored the family composition and socioeconomic position of these children, and found that children with LLC were more likely to live in a family where both parents were unemployed compared to the non-LLC group (33% vs 20%), and in a single-parent household (29% vs 20%). However, there were no differences between the two groups in terms of material hardship, area deprivation, maternal educational qualifications, family size, and whether children's parents were more likely to be married then cohabiting when living together (Project 2).

Measures

1. Children's LLC status and developmental outcomes

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In MCS3, mothers were asked to identify any long-standing illnesses the child may have had, and these were coded (post-interview; up to six illnesses) on the basis of ICD-10 (NatCen, 2006). ICD-10 data in the MCS3 were compared against the ICD-10 codes on life-limiting conditions in the Hain dictionary (Hain et al., under review), a published list of diagnoses and codes drawn from death data and referrals to clinical paediatric palliative care services. Codes were merged to identify children with *any* life-limiting condition among reported long-standing illnesses. Among the children with a life-limiting condition, 93.78% had one LLC, whilst 6.22% had two LLCs.

In the present study, we examined children's cognitive and behavioural development, and their physical health status. Cognitive development was captured using three subscales (picture similarities, naming vocabulary, and pattern construction) of the British Ability Scales (BAS-II; Elliott, Smith, & McCulloch, 1996). The BAS-II is a standardised measure of intellectual functioning frequently used in clinical, educational practice and research. In MCS3, scores were subjected to a principal components analysis, which confirmed the presence of a factor *g* as an index of general cognitive ability, accounting for about 56% of the overall variance (Jones & Schoon, 2008; Schoon, Cheng, & Jones, 2010). A presence of an intellectual disability (ID) was indicated by a score at or below two standard deviations of the standardised *g* scores (cf., Totsika et al., 2011). This indicated a 3.1% weighted prevalence of ID overall among MCS3 singleton children.

The Strengths and Difficulties Questionnaire (SDQ; Goodman, 1997) measured behavioural and emotional problems at age 5. Due to its ease of use and excellent psychometric properties (Goodman, 2001), the SDQ is frequently used in research and clinical practice to assess behaviour problems and indicate potential clinical levels of difficulties. It measures hyperactivity, emotional symptoms, conduct problems, peer relationship problems and prosocial behaviour. A total behaviour problems score is obtained by combining the first

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four subscales (range 0-40; Cronbach's alpha for these data: .80). Available cut-off scores indicate borderline and abnormal levels of problem behaviours (http://www.sdqinfo.com), and were used in the present study to indicate rates of caseness (borderline and abnormal levels of behaviour problems).

MCS3 interviewers obtained measurements of children's height and weight used to calculate Body Mass Index (BMI). Using age and gender-specific cut-offs, MCS3 estimated obesity levels (Griffiths et al., 2010). Among the 15,246 first cohort children in MCS3, about 5.5% had a BMI which placed them in the obese range (weighted). Data from other Western countries such as the US (Ogden and Carroll 2010) suggest this may underestimate population prevalence.

2. Parental well-being

Information was obtained through interviews with the child's main carer using Computer Assisted Personal Interviewing (CAPI). In 97.14% of cases the main carer was the child's mother (N= 14,810; 396 were male relatives and 40 were another female relative). As only a small proportion of interviews were conducted with non-maternal carers, these were retained in the analyses, and for ease of reference parental outcomes will be referred to as maternal.

Psychological distress was measured using the K6 (Kessler et al., 2002). The K6 is widely used to screen community populations for psychiatric disorders. It includes six items on symptoms present over the past 30 days, which can be summed into a total K6 score (range 0-24) to indicate high levels of psychological distress (α:.88). A cut-off is also available to predict serious mental illness (SMI: at least one 12-month DSM disorder, other than substance use; Kessler et al., 2003).

Physical health was reported by mothers as excellent, good, good, fair or poor in answer to the question 'How would you describe your health'? Similarly, a 1-10

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point item rated overall satisfaction with life so far, with higher values indicating higher satisfaction. Satisfaction with partner relationship was assessed using four items from the Golombok Rust Inventory of Marital Satisfaction (GRIMS: Rust et al., 1990). They were grouped to indicate overall relationship satisfaction (range 4-20, α: .81), with higher scores suggesting *lower* satisfaction with partner relationship.

3. Family socioeconomic position

A robust index of socioeconomic position (SEP) was created using a variety of economic indicators. Household work status (neither parent working vs at least one working), area deprivation (area in the lowest 20% of the Index of Multiple Deprivation [combining area information on income, education, housing, crime, employment, and health]), relative poverty (inability to afford at least two out of five socio-culturally defined standards of living [e.g., a weatherproof coat for the child]), and poverty (income <60 equivilised median income for the UK) were combined to describe the family's SEP (α: .67), with higher values indicating higher levels of deprivation.

Statistical analysis

MCS data were obtained from the UK Data Archive (http://www.data-archive.ac.uk/). The ethical responsibilities of the present authors included the protection of participants' anonymity and confidentiality. Because of the complex design of the MCS, weights were used throughout all analyses to account for both the disproportionate sampling of particular groups and nonresponse/attrition rates. All analyses were conducted in STATA 11 (StataCorp., 2009) using the survey data analysis environment.

To address the first two research questions, we adopted a two-group design comparing outcomes for children with and without LLC. Where continuous

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measures had available cut-offs (to indicate clinical caseness as described in Measures above) comparisons were done on both types of outcomes. We estimated the standardised mean difference (*d*) and relative risk (RR) as effect sizes for continuous and binary outcomes, respectively. As with all analyses, effect sizes were estimated on weighted data. For the first research question, we compared levels of intellectual disability, obesity and behaviour problems between the two groups of children. For the second research question, we compared the two groups of mothers on their self-reported health status, psychological distress (and levels of SMI caseness), life satisfaction and satisfaction with partner relationship.

To explore the association between LLC and children's development (third research question), we adopted two modelling approaches. First, we wanted to see whether LLC is significantly associated with development, after controlling for the effect of other important variables. We fitted multiple regression models in the overall MCS sample, controlling for the effects of intellectual disability, child gender, maternal psychological distress, family socioeconomic position, and maternal physical health. Two such models were fitted, one for children's total behaviour problems, and one for BMI. Where the effect of LLC presence was significant, further logistic regression models were conducted within the LLC sample. The goal of these models was to examine the separate effect of each predictor (child gender, intellectual disability, family SEP, maternal psychological distress and self-reported physical health) on caseness for obesity and behaviour problems.

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3.3 Results

Comparisons between children with LLC and those without

The upper part of Table 3.1 presents children's scores on total behaviour difficulties, BMI and levels of intellectual disability, obesity, and

borderline/abnormal behaviour problems. Comparisons between children with LLC and the comparison group without LLC suggested all differences were significant, with a moderate sized difference for total behaviour problems (d=0.57, 95% CIs 0.46 to 0.69, p<.001), and a small sized difference for BMI scores (d=0.16, 95% CIs 0.04 to 0.28, p=0.009). Children with LLC had higher levels of behaviour problems and higher BMI scores. The caseness comparisons suggested that children with LLC were about 6 times more likely to present borderline or abnormal levels of behaviour problems (95% CIs of RR 4.56 to 8.45), and about 2.5 times more likely to have a BMI in the obese range (95% CIs of RR 1.63 to 4.10). They were 11 times more likely than their non-LLC peers to have an intellectual disability (95% CIs of RR 7.55 to 15.98).

	LLC group	Non LLC group	Difference[1]	Effect size[2] (95% CIs)
CHILDREN				
Total Behavioural difficulties (SDQ)(SE)	12.47 (.54)	7.19 (.07)	t= 9.80, p<.001	d=0.57 (.46 to .69)
BMI (SE)	16.73 (.15)	16.34 (.02)	t=2.62, p=0.009	d=0.16 (.04 to .28)
% Borderline/Abnormal behaviour problems	42.72%	10.72%	F=174.02, p<.001	RR: 6.21 (4.56 to 8.45)
% Intellectual disability	23.28%	2.69%	F=243.21, p<.001	RR: 10.98 (7.55 to 15.98)
% Obese	12.78%	5.36%	F=17.64, p<.001	RR: 2.59 (1.63 to 4.10)
MOTHERS				
Self-reported health status (SE)	2.60 (.09)	2.39 (.02)	t=2.38, p=0.018	d=0.14 (.02 to .25)
Psychological distress (K6) (SE)	4.19 (.29)	3.11 (.04)	t=3.79, p<.001	d=0.22 (.11 to .34)
Life satisfaction (SE)	7.20 (.15)	7.49 (.02)	t=1.88, p=0.061	d=0.11 (-.01 to .22)
Satisfaction with relationship (GRIMS) (SE)	15.43 (.25)	16.07 (.04)	t=2.54, p=0.012	d=0.18 (.04 to .31)
% Serious Mental Illness (K6)	4.79%	3.38%	F=0.140, p=0.237	RR: 1.44 (.78 to 2.63)

[1]groups[1] T-test for continuous outcomes; Chi-square for binary outcomes, corrected for survey design (F; Rao & Scott, 1984).) [2]d: standardised mean difference for continuous outcomes; RR: Relative Risk for binary outcomes

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Table 3.1: Comparison of child developmental outcomes and maternal well-being between the LLC and non-LLC groups

The lower part of Table 3.1 presents mothers' scores on self-rated health status, psychological distress, life satisfaction, relationship satisfaction and levels of likely psychiatric disorder. Comparisons between the LLC and non-LLC groups suggested that LLC mothers reported slightly worse physical health (d= .14, 95% CIs .02 to .25; p=0.018), and slightly elevated psychological distress (d =0.22, 95 CIs .11 to .34; p<.001). However, it is important to note that these group differences are only small in size and may not be clinically meaningful. Furthermore, mothers in the LLC group were not more likely to score above cut-off for a likely psychiatric disorder (SMI RR 1.44, 95% CIs .78 to 2.63). In addition, they were as satisfied with their life as mothers of children without LLC (d=0.11, 95% CIs -.01 to .22, p= ns), and they were actually slightly more satisfied with their partner relationship than comparison mothers (d=0.18, 95% CIs .04 to .31).

The association between LLC and problem behaviours and BMI

Controlling for any potential effects of intellectual disability, child gender, maternal psychological distress, family SEP and maternal physical health, the presence of LLC was significantly associated with children's total SDQ scores (beta: .10, p<.001). Similarly, after controlling for the same factors, the presence of LLC was associated with children's BMI scores, albeit marginally (beta: .02, p=0.048). Table 3.2 presents the results of these models.

	SDQ total behaviour problems	BMI scores
	N_w=14,215	N_w=14,210
Model fit	R^2 =0.215, p<.001	R^2 =0.005, p<.001
Child gender	-.09, p<.001	-.03, p=0.002

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Intellectual disability	.12, p<.001	.04, p=0.018
Family SEP	.17, p.<001	.03, p=0.003
Maternal psychological distress (K6 total)	.28, p<.001	.002, p=0.865
Maternal physical health	.09, p<001	.02, p=0.018
Life-limiting condition	.10, p<.001	.02, p=0.048

Table 3.2. Association of LLC with behaviour problems and BMI scores. Fully standardised betas from multiple regression models are presented in cells.

Factors associated with elevated behaviour problems and obesity in children with LLC

As the previous models suggested that LLC was significantly associated with children's behaviour problems and BMI, we explored the contribution of each factor to obesity and borderline/abnormal problems of problem behaviour within the LLC group only. The focus here was to explore the adjusted relative risk associated with different family factors within this group of children, given that they are more likely to present high levels of behaviour problems and obesity levels. Table 3.3 presents the results of the multivariate logistic models. High levels of problem behaviours were predicted significantly by all factors with the highest relative risk associated with the presence of an intellectual disability (RR_{adj}: 5.14, 95% CIs 1.99 to 13.25), followed by poorer reported maternal physical health (1.63, CIs 1.15 to 2.32), family SEP (1.50, CIs 1.06 to 1.29), and maternal psychological distress (1.15, CIs 1.03 to 1.29). Obesity in children with LLC was not really associated with any of these factors, with the exception of family SEP (RR_{adj}: 1.82, CIs 1.01 to 3.28).

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	SDQ total behaviour problems	BMI scores
	N_w=256	N_w=247
Model fit	F =8.39, p<.001	F=3.48, p<.004
Child gender	.20 (.08 to .49)	.37 (.11 to 1.23)
Intellectual disability	5.14 (1.99 to 13.25)	1.98 (.59 to 6.60)
Family SEP	1.50 (1.06 to 1.29)	1.82 (1.01 to 3.28)
Maternal psychological distress	1.15 (1.03 to 1.29)	1.07 (.97 to 1.19)

(K6 total)		
Maternal physical health	1.63 (1.15 to 2.32)	.97 (.52 to 1.83)
Life-limiting condition	5.14 (1.99 to 13.25)	.37 (.11 to 1.23)

Table 3.3. Risk for high levels of behaviour problems and obesity in children with LLC. Adjusted relative risks (and 95% CIs) from logistic regression models are presented in cells.

3.4 Discussion

The aim of this study was to explore the development of children with a life-limiting condition, and the psychological and physical wellbeing of their mothers, and through these to examine the putative impact on a child's development of having a life-limiting condition. For the first time, we were able to interrogate an existing cohort database in order to compare data in the two groups. This was made possible by the development of a 'dictionary' of ICD 10 codes based on the ACT/RCPCH criteria for life-limiting conditions Hain et al, under review, (Fraser, Miller et al. 2012). This approach avoided the risk of recruitment bias and allowed us to study a numerically large group, something that is otherwise difficult in children's palliative care.

In considering the impact of a LLC on the child's development, the most striking difference between the two groups was the risk of high levels of behaviour problems (table 3.1). Compared with children without LLC, the relative risk of intellectual disability was 10.98 (range 7.55-15.98). Behavioural difficulty is known to be associated with intellectual disability (Einfeld, Tonge et al. 2006; Forster, Gray et al. 2011; Gray, Piccinin et al. 2011). The importance of behavioural symptoms in our study, rather than physical ones, confirms other published findings in families of children with LLC (Wood, Simpson et al. 2010; Malcolm, Forbat et al. 2011).

The presence of cognitive impairment in the child remained the most powerful association with behavioural difficulties among families of children with LLC,

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though there were numerous other factors associated with increased risk (table 3.3). These included the child's gender, family socioeconomic status, maternal psychological distress and maternal general health. The association was illustrated in our study, not only by how likely a given child is to demonstrate behavioural difficulty, but also how severe that behavioural difficulty is likely to be. This concept of 'caseness' here reflects children who are likely to have clinical levels of behavioural difficulties.

An interesting observation is that obesity was much more likely among children with life-limiting conditions. The association of obesity and life-limiting conditions seems to be independent of related variables such as intellectual disability, maternal psychological distress and family socioeconomic position. Of these, it was only family SEP in our study that impacted significantly on the likelihood of obesity within the group of families of children with LLC. This may be at least partly related to parental BMI, which was not recorded in our study.

It seems likely that there is a two-way association between obesity and functional impairment (Heo, Pietrobelli et al. 2010). Obesity also makes pain due to musculoskeletal causes more likely (Pain and Wiles 2006; Rimmer, Rowland et al. 2007). Furthermore, there is evidence that the number of children living with life-limiting conditions is rising (Taylor, Miller et al. 2010) so that the long-term health consequences of obesity, such as hypertension and diabetes, may become increasingly problematic. The practical impact of obesity in a child with limited mobility is significant for those caring for the child, particularly if there is only one parent.

Among mothers of children with life-limiting conditions, general health status was less good, and psychological distress was higher. The use of a single self-report item on physical health status, though less robust than a health-related quality of life measure, was sufficiently sensitive to identify differences between the two groups of mothers in this respect.

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Despite the abundance of causes of distress, the evidence from our study is that this did not increase the likelihood of serious psychopathology (table 3.1) in mothers when their child with LLC was still young. Perhaps more surprisingly, mothers in the families of children with LLC group did not seem to be any less satisfied with their life than those without. Rather the reverse; there was a trend towards higher life satisfaction among families of children with LLC. This apparently unexpected observation in fact supports the conclusions of other studies that families caring for children with complex chronic conditions are often resilient and can indeed describe many positive outcomes of their experience (Smith, Oliver et al. 2001; Gardner and Harmon 2002; Levine 2009; Wood, Simpson et al. 2010). This supports the findings of other work in mothers of children with developmental disabilities (Hastings & Taunt, 2002) and autism (Totsika et al., 2011) that suggest preservation of the potential for positive psychological adaptation, and that mothers of children with disabilities can simultaneously experience impacts on mental health that are both positive and negative. Further evidence of this was provided by our finding (table 33.1) that mothers of children with LLC were more satisfied with relationships with their partner. It may be that working together to care for their child allows parents to become more effective in their communication and to develop a stronger partnership (Hobson and Noyes 2011).

3.5 Conclusion

In summary, our results seem to indicate that children with life-limiting conditions are more likely than other children to have an intellectual disability and high levels of behavioural difficulties, and that this causes considerable distress to their families. It further suggests that families are often already coping with poor socioeconomic status, low income and single parents. Despite the distress that this causes, however, our study demonstrates that families remain remarkably resilient in the face of this onslaught. There is nothing to suggest the risk of

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serious mental illness is increased by having to care for such a child, nor even that satisfaction with life is necessarily impaired by it. Consistent with Aristotle's conception of well-being, the results from our study suggest that satisfaction with partner relationships might even be increased by the task.

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Project 4:

Psychological burden of life-limiting conditions in childhood: A brief integrative review of concepts, definitions and epidemiology.

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Executive summary of project 4

Our aim:
To identify current concepts and definitions, epidemiology and psychological problems in UK Children's Palliative Care within the UK.

What we did:
We reviewed the published literature, using as a guide three questions:

- How is children's palliative care defined and conceptualised in policy and literature?
- What does the literature tell us about the numbers and characteristics of children requiring palliative care?
- What psychological problems are associated with palliative care needs in children?

Headline findings:
- The ACT (2009) definition is widely recognised and used in policy documents in England and Wales.
- Data on the numbers of children requiring palliative care in the UK at any one time is based on estimated figures derived from a number of different sources.
- Children, young people and their families requiring palliative care have a broad range of characteristics and needs requiring flexible services.
- Psychological problems significant for many children and families with LLC

Comments:
This review was conceived as a support for the other three projects, as we uncovered ambiguities in the definition of key terms, inconsistent empirical evidence, and diversity among the sources of published data in respect of psychological problems.

Status:
This project is complete and a manuscript prepared. Its main function is as a resource, but we anticipate also extending the report on the review and exploring publication being able to publish it as a separate paper.

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4.1 Introduction

The purpose of this integrative review is to explore the concepts, definitions, epidemiology and psychological problems encountered within Children's Palliative Care in the UK. Three key questions guided the review process:

- How is children's palliative care defined and conceptualised in policy and literature?
- What does the literature tell us about the numbers and characteristics of children requiring palliative care?
- What psychological problems are associated with palliative care needs in children?

> *"Palliative care for children and young people with life limiting conditions is an active and total approach to care, from the point of diagnosis or recognition, embracing physical, emotional, social and spiritual elements through to death and beyond. It focuses on enhancement of quality of life for the child or young person and support for the family and includes the management of distressing symptoms, provision of short breaks and care through death and bereavement."* (ACT 2009)

The terms "life threatening" and "life limiting" are often used interchangeably when describing a child or young person who may be in need of a palliative care service but in the context of this review the classification of these conditions is defined as follows.

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<u>Life-limiting/life-shortening conditions</u>

Life-limiting /life-shortening conditions are those for which there is no reasonable hope of cure and from which children or young people will die. Some of these conditions cause progressive deterioration rendering the child increasingly dependent on parents and carers.

Life-threatening conditions

Life-threatening conditions are those for which curative treatment may be feasible but can fail, such as cancer. Children in long-term remission or following successful curative treatment are not included.
(ACT 2009)

Similarly child, young person and young adult have moving boundaries in terms of age classification years. We have used the following to guide this review.

Child: A child is defined as a young person aged up to their 19th birthday.

Young adult: The term young adult describes a person from their 19th birthday.

Young person: The term young person describes a person from their 13th – 19th birthday.

Furthermore four broad categories of life-threatening and life-limiting conditions may be delineated.

Category 1 Life-threatening conditions for which curative treatment may be feasible but can fail. Where access to palliative care services may be necessary when treatment fails or during an acute crisis, irrespective of the duration of that threat to life. On reaching long-term remission or following successful curative treatment there is no longer a need for palliative care services. Examples: cancer, irreversible organ failures of heart, liver, kidney.
Category 2 Conditions where premature death is inevitable, where there may be long periods of intensive treatment aimed at prolonging life and allowing participation in normal activities. Examples: cystic fibrosis, Duchenne muscular dystrophy.
Category 3 Progressive conditions without curative treatment options, where treatment is exclusively palliative and may commonly extend over many years.

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Examples: Batten disease, mucopolysaccharidoses.
Category **4** Irreversible but non-progressive conditions causing severe disability leading to susceptibility to health complications and likelihood of premature death. Examples: severe cerebral palsy, multiple disabilities such as following brain or spinal cord injury, complex health care needs and a high risk of an unpredictable life-threatening event or episode.

(ACT 2009)

4.2 Methods

- Rationale for methods

The integrative literature review method was chosen to allow for the inclusion of diverse methodologies and to allow a varied sampling frame.

It follows the five stages that are described by Whittemore & Knafl (2005): problem identification, literature search, data evaluation, data analysis and presentation. 3 individual literature searches were undertaken to address each question separately.

- Search strategy

A search protocol was developed to inform the search strategy and outline search criteria and terms. Search terms were different for each question. The separate searches had different inclusion and exclusion criteria. This is summarised in Table 4.1 below. A computerised search was then conducted to identify relevant studies. Multiple databases were identified and searched including CINAHL, MEDLINE, Google Scholar, The Cochrane Library , ASSIA: Applied Social Sciences Index and Abstracts, PILOTS Database, PsycINFO, British Medical Journal Publications, Association for Children's Palliative Care (ACT) Paed Pall Lit, Journals at OVID and Royal College of Nursing Publications.

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For question 1 a manual search was done online, searching all UK Government departments, executive agencies and non-departmental public bodies using the search terms “child” and “palliat*”. All those were retrieved with a relevant policy definition. Cross referencing was then used from one policy to another to check inclusions of other policies.

A further search was done of the 4 UK and 1 International agencies concerned with palliative care for their definitions: National Council, Association for Children’s Palliative Care (ACT), Children’s Hospices UK, Help the Hospices and the World Health Organisation.

For question 2, condition specific searches were also undertaken for 22 of the most frequently occurring International Classification of Diseases (ICD10 World Health Organisation (WHO) conditions which have been judged by professionals working in paediatric palliative care to be life limiting (Hain et al 2010) using keywords such as Cerebral Palsy, Muscular Dystrophy, Spinal Muscular Atrophy, Leukaemia. Disease specific National Registries were also searched and specific charity websites (subject to existence) for each of the 22 conditions mentioned above. Data was extracted using a specifically developed extraction template. Quality appraisal of the data was not within the remit of this integrative review.

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Question	**Setting**	**Perspective**	**Intervention /issue of interest**	**Evidence**	**Keywords/ Search terms**
1/How is children’s palliative care defined and conceptualised in policy and literature?	UK only Secondary care, tertiary care, children’s hospice, hospice at home service, school, community, primary care, third sector	Multiple perspectives- professional, patient and family	Children’s palliative care age 0-25/16-30	UK policy, practice and grey literature **1990** onwards	Child* Paed* Palliat* Care Definitions Concepts
2/What does the literature tell us about the numbers	UK only Secondary care, tertiary care,	Epidemiology	Numbers and characteristics of	UK reports, audits, evaluation	Child* Paed* Palliat* Care

and characteristics of children requiring palliative care?	children's hospice, hospice at home service, school, community, primary care, third sector		children age 0-25/16.30	data and studies since **2005**	Numbers, Epidemiology, Data, Statistics Prevalence, Characteristics, Incidence Condition specific searches as per Hain et al (2010)
3/ What psychological problems are associated with palliative care needs in children?	UK and International Secondary care, tertiary care, children's hospice, hospice at home service, school, community, primary care, third sector.	Multiple perspectives- patient/family / extended family (exclude psychological impact on professionals)	Psychological impact, implications of condition, impact of care and services	UK and international policy, practice and grey literature **1990** onwards English language only	Child* Paed* Palliat* Care Psychology Mental health Siblings Paternal Maternal Psychological problems Impact Wellbeing

Table 4.1

4.3.Results

- Retrieval of references

Searches for questions 2 and 3 followed the same format. In the first phase a computerised search was done for each search question and the abstract obtained. In phase two abstracts were scrutinised according to the inclusion, exclusion criteria and test of relevance. Full papers were then retrieved and read and any duplicates removed. A total of 159 papers were included in the review.

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Table 4.2 summarises the results for each search question.

Question	Abstracts retrieved	Full papers retrieved
1/How is children's palliative care defined and conceptualised in policy and literature?	24	24
2/What does the literature tell us about the numbers and characteristics of children requiring palliative care?	107	43
3/ What psychological problems are associated with palliative care needs in children?	186	92

Table 4.2

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Findings in respect of specific questions

Question one: How is children's palliative care defined and conceptualised in policy and literature?

Children's palliative care is provided by a range of different providers within both the statutory and voluntary sector, with the Children's National Service Framework (DH 2004, 2006, 2007) defining the overall health and social care service standards within the UK. Specific to Children's Palliative Care "Better Care, Better Lives" (DH 2008) provides best practice guidance for the sector.

The first national definition of children's palliative care evolved in 1993. Developed by the Association for Children's Palliative Care and the Royal College of Paediatrics and Child Health it has subsequently been reviewed but in its most recent version (ACT 2009) its core original elements remain unchanged. The only adaption has been to emphasise that palliative care should be from the point of diagnosis or recognition of a life limiting condition and that short breaks as opposed to respite are provided. These changes were made during the same

year as the Disabled Children's sector became more closely involved with the Children's Palliative Care sector (DH/DCSF 2008) as it was recognised that there is an overlap between the needs of children and young people requiring palliative care and those living with disabilities and other complex care needs. (Craft and Killen 2007)

Differences were found to exist with the universal application of the ACT definition within each individual countries policy context. Within the UK for the period covered by this review Scotland was found to be utilising the World Health Organisation definition (NHS Scotland 2008) and Northern Ireland has its own individual definition (DHSSPS 2007). No evidence was found for the Republic of Ireland. Within England the most recent policy documents specific to children's palliative care (2005 onwards) utilise the ACT definition, earlier documents such as some of the National Service Framework documents refer to long term conditions, severe disability or high care needs. Cancer policy documents similarly use their own definition of palliative care. More generic children's policy documents appear to not define children's palliative care but use the terms life limiting, life threatening, disability, vulnerable child, long term conditions and complex health care needs.

Of the 24 reports included, 8 key concepts were elicited which underpin the nature of children's palliative care.

- Holistic support (type): psychological, spiritual, physical, emotional, social, psychosocial, practical, functional.
- Holistic support (person): child, family, siblings, immediate and extended family, schools and staff.
- Support through illness trajectory from diagnosis through death, bereavement and beyond.

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- Choice about location of and type of care provision and treatment: day care, hospice at home, hospice, hospital, community, specialist respite, telephone/advice, bereavement support, equipment provision.
- Care types: Pain and symptom management, terminal care, end of life, emergency care, contact/key worker visits, respite/short breaks, counselling, pre and post bereavement care, specialist palliative care, general palliative care**,** complimentary therapies, long term.
- Focus on life and its quality, independence and transitions.
- Recipients of care: Complex care needs, child with disability, clinically dependent.
- Method of delivery: multiagency assessment, integrated, culturally and spiritually sensitive, individualised, accessible, 24hrs, specialist or generalist.

Care for the whole person within its many facets is emphasised as is the importance of this care being delivered to not just the life limited child or young person, but for the whole family including grandparents and also those who may be part of a much wider network of contacts that the child or young person may have in their life. The support offered is provided over a broad timescale but can range from a number of days to many years depending on the diagnosis of the sick child and delivered within a range of locations. The type of care provided can vary dependent on the need and the individual choice and wishes of the child/young person and family. However, the provision of support so that the child and family can remain as independent as possible and with as much quality of life as possible is paramount. Distinctions are made between what is generic or specialist palliative care and which children may be in need of palliative care provision. (DH 2005)

Question 2: What does the literature tell us about the numbers and characteristics of children requiring palliative care?

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From 2004-2008 the UK had a focus on developing policy and practice for children with complex health care needs. This was instigated by the House of Commons Report into Palliative Care in 2004, and was then followed by a National Service Framework (DH/DFES 2004), Commissioning Guide (DFES/DH 2005), and Best Practice document (DH 2008). During this time there was also an Independent Review (Craft and Killen 2007) and an Economic Study (Lowson et al 2007) to support the Independent Review. The Independent Review highlighted that evidence on the numbers of children with life limiting conditions is imprecise and utilised a number of different sources to identify a method by which an economic study could be based. Lowson et al (2007) also found wide variation in the definitions of children with disabilities, with complex needs and requiring palliative care and identified that Primary Care Trusts (PCTs) had problems identifying these children and therefore providing appropriate palliative care services.

The method used was one previously used by Parker et al (2003) where the number of children with disabilities was estimated from literature sources, disease registers, UK census data and OPCS estimates for a population of 250,000. Parker et al (2003) utilised this approach for a typical population of 50,000 children to identify a "best estimate". This best estimate identified the following number of cases for 0-14 year olds in a typical locality of 50,000 children, for children with a life limiting condition shown in Table 3.

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Nature of disability	**No of cases**
Muscular	8
Cystic Fibrosis	21
Fragile X	8
Juvenile chronic arthritis (proxy for other LLCs)	50
Technology dependent	25
Complex needs	1,500
Cerebral palsy	110

Table 4.3

It is not possible from the figures above easily to identify similarities and differences when compared to the findings identified from the 43 papers/sources of evidence. This is for a number of reasons. The first is that the studies included have varying geographical boundaries for the populations they have identified which range from countries, health districts, regions or by Strategic Health Authority (SHA). Secondly studies vary in their age banding. Thirdly the sources of data vary wildly in the formula and approach that they have used to establish incidence or prevalence.

Some studies however are able to identify an actual change in prevalence (Odding et al 2006), whilst others demonstrate possible changes by using estimated prevalence based on analysis of mortality data and HES data (Baker et al 2010). Other studies identify the numbers of children with complex needs based on the numbers of those claiming higher rate Disability Living Allowance but are unable to identify the characteristics of those needs. (Nef 2009)

In 2007, following the Independent Review, a revised formula was established to estimate the numbers of children requiring palliative care. This was based on a study by Cochrane et al (2007) who conducted an analysis of mortality data and hospital admission data, for children and young adults with conditions likely to require palliative care services in England. This was estimated as 15 per 10,000 (excluding neonates) and 16 per 10,000 (including neonates) which equated as 20,100 children and young people living with conditions likely to require palliative care in England.

Of the 43 papers/sources of evidence which fulfilled the inclusion criteria for this question, 5 utilised or citied this formula for calculating prevalence. 3 of these sources of evidence were government reports, the other 2 were reports for Children's Palliative Care Networks. Other methods citied included examining

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condition specific registry data, identifying literature reviews of previous data sources, analysis of death certificate data, analysis of Hospital Episode Statistics (HES) data, secondary data analysis, examination of specific datasets, and using estimates based on others citied estimates. The papers/sources could also be classified by condition. The following groups of conditions were identified: cardiac tumours, cerebral palsy, cancer, cystic fibrosis, deaths of children occurring at home, deaths of children occurring in hospitalised children, disabled children, epilepsy, heart failure due to heart muscle disease, disabled children with complex needs, genetic muscle disease, inherited metabolic disorder, leukaemia, long term ventilation, mucopolysaccharidosis, muscular dystrophy, neuromuscular conditions, palliative care, palliative care needs, renal replacement therapy, technology assisted children and young people and terminal conditions.

The needs of the children, young people and their families contained within the papers were identified. These ranged from being technology dependent and requiring interventions such as tracheostomy care, oxygen therapy, parenteral and enteral feeding, peritoneal dialysis, IV drug therapy, non invasive ventilation, (Kirk 2008), mechanical ventilation, ECMO, cardiac transplantation, drug therapy, arrhythmia therapy (Andrews et al 2008) to being unable to walk and having a sensory impairment or intellectual impairment (Mongan et al 2006). Depression, panic disorders or anxiety were also identified as mental health problems that will affect a third of young adults with epilepsy. (Beecham et al 2010)

Craft & Killen (2007) also highlighted that children and young people are living longer with palliative care needs and the increasing complexity of these.

One study identified that Infants with congenital malformations or perinatal conditions were more likely to die in ICU and older children with malignancy outside the ICU. There were a greater proportion of children having end of life events managed on ICU. (Ramnarayan et al 2007) Similar findings were

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established by Fraser et al (2010) who found that children admitted to Paediatric Intensive Care Units (PICU) had a very low rate of discharge to palliative care.

Question three: What psychological problems are associated with palliative care needs in children?

The 92 papers included in the review literature were collated and organised according to which family member was studied, medical condition/diagnosis of the sick child or young person, age of the child/young person and psychological problem identified.

The psychological problems identified in the literature were as follows:

- Distress
- Conduct, hyperactivity, emotion and peer problems
- Psychological problems related to functional ability and pain
- Depression
- Low mood states,
- Fear
- Withdrawal
- Unfavourable behavioural and emotional outcomes
- Stress
- Mental disability
- Impaired concentration
- Anxiety
- Problem behaviours
- Sleeping difficulties
- Anticipatory grief
- Psychiatric symptoms
- Posttraumatic stress
- Learning difficulties
- Psychological suffering
- Acute stress disorder
- Chronic grief
- Chronic sorrow
- Recurring grief
- Isolation
- Anxiety attacks
- Hopelessness and helplessness
- Constant worry
- Increased alcohol and drug use
- Suicidal thoughts, attempted suicide
- Nervous breakdown
- Affective disorders
- Schizophrenia
- Substance misuse
- Loneliness
- Emotional responses: frustration, sadness, jealousy, regret, anger, denial, despair

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Of the 92 papers, 41 used Cancer as the focus population for study. Cancer in children was associated with isolation causing distress (Enskar at el 2008), low mood (Chao et al 2003), fear (Carlsson et al 2008), depression, withdrawal and social problems (Poggi et al 2005), and distress (Trask et al 2003).

Psychological problems were seen to impact on the whole family: the sick child, mothers, fathers, grandparents, and siblings and across a wide range of conditions and stages including: Cancer, Cerebral Palsy, being ventilator dependent, HIV, Cardiac surgery and heart disease, Leukaemia, chronic illness, palliative care or end of life care, Cystic Fibrosis, Neuromuscular disease, Juvenile Huntingtons Disease, and Neurodevelopmental and Neurodegenerative disorders.

12 studies also looked at the impact of the death of a child. The majority of the studies focus was either on the mother, father or sick child. Only a few studies focused on grandparents or siblings. 4 studies focused exclusively on the experience of fathers.

Some emerging elements which featured in a number of studies were concerned with the level of psychological impact and adjustment and the distance in terms of time from when the diagnosis was made, the use of cognitive coping strategies, and the levels of professional and non professional support and subsequent adjustment of the family.

The literature which explored the concept of fear could also be further classified according to its cause. Fear could be related to the physical body and self image or could be experienced because of actual or anticipatory loss and therefore fear of separation. Fear could be experienced because of episodes of crisis, or as a reaction related to a diagnosis.

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The death of a child was associated with increased risk of or actual anxiety or depression (Aho 2006, Younblut 2010, Jiong et al 2005, Barrera et al 2009, Lannen et al 2008, Goodenough et al 2004, Kreicsbergs et al 2004) and risk of psychiatric hospitalisation increased (Jiong et al 2005)
Furthermore Kersun et al (2007) suggested that symptoms of anxiety and depression are under recognised and under treated in children at the end of life.

Parental distress was found to be not strongly related to the child's illness severity (Thompson et al 1992, Davis et al 1998, Nereo et al 2003, Uzark 2003)) but is found to be higher at the point of time around diagnosis (Vrijmoet-Wiersma 2008, Wijnberg-Williams 2006, George et al 2006). Poor maternal adjustment was associated with higher levels of daily stress (Thompson et al 1994, Han 2003)

Some researchers have found a direct relationship between levels of support received and subsequent emotional impact and levels of stress experienced (Wong et al 2008, Lach et al 2009) however in contrast others have found that social support had a negligible effect on mothers psychological functioning. (Wijnberg-Williams 2006)

Some studies explored transaction stress and coping models and looked at adaptive coping (problem focused, cognitive restructuring, seeking information, seeking social support) and palliative coping (emotion focused, avoidance, wishful thinking, self blame).

Passive reaction patterns were correlated with higher psychological distress (Stam et al 2008) and adaptive style was significantly associated with posttraumatic stress symtomatology in children with cancer and their parents (Phipps et al 2005)

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High levels of parent perceived stress, high parent use of palliative coping methods, relative to the use of adaptive coping and high levels of family conflict were associated with poor psychological adjustment in parent and child adjustment to Duchenne Muscular Dystrophy. (Thompson et al 1992) Self blame and behavioural disengagement were coping strategies associated with child and parent maladjustment (Wong et al 2008) Active coping (planning, taking direct action and increasing coping efforts) in bereaved mothers was associated with less intense grief reactions. (Riley et al 2007) Post traumatic stress disorder/post traumatic stress symptoms were found to decrease over time from diagnosis (Jurbergs et al 2009, Poder et al 2008) however PTSD in life limiting paediatric illness can be reduced or prevented and treated when it occurs. (Stuber et al 2006) Siblings who receive low levels of social support were also found to be at risk for psychological maladjustment (Barrera et al 2004) and would benefit from opportunities to meet with health care providers to discuss their feelings associated with having a brother or sister with cancer (Packman et al 2008)

Implications for practice

Some of the findings suggested ways in which the psychological problems could be addressed. These are broken down by family member.

Interventions for the child

These ranged from the importance of understanding that increased distress in children was related to the treatment period and for a need for greater awareness amongst nursing staff (Enskar et al 2008), greater awareness of the potential for psychological problems in children with cerebral palsy (Parkes et al 2008), the importance of the mental health team for children who are ventilator dependent (Sarvey 2008), the importance of open communication (Battles et al 2002), teaching coping skills as a means of preventative interventions for psychological adjustment (Meijer et al 2002, Miller et al 2009), and the provision of psychotherapy may facilitate psychological adjustment (Brown et al 2006)

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Interventions for the mother

Professionals should be aware that long term maternal distress can have an influence on parent reported problems in children (Spijkerboer 2010) The need was identified for individual assessment of families and focused intervention in managing stress levels (Glenn 2008) and for increased services to help with families cope with the effects of Duchenne Muscular Dystrophy (Nereo 2003) and neurodevelopmental disorders (Lach et al 2009).

Interventions for fathers

Recognition is needed that fathers also need support in expressing and managing their grief (Aho 2006), with services specifically designed to meet the psychological needs of fathers as well as mothers (Ware et al 2007)

Interventions for parents

Clinicians must assess parenting stress to provide appropriate support and guidance (Uzark 2003), especially during the diagnosis phase (Piersol et al 2008)

Interventions for siblings

Healthcare professionals have an important role in supporting parents but also in encouraging them to involve the siblings in their brother or sisters illness (Kreicbergs 2010)

Interventions for grandparents

Pediatric nursing practice needs to acknowledge and add grandmothers' experience to it's nursing knowledge (Hall et al 2004) and to also provide support to this group. (Youngblut et al 2010)

Interventions for the child and parent

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Those interventions that aim to enhance vicarious hope (expectation that desirable things will occur in the future) and reduce vicarious despair (expectation that undesirable things will occur in the future) can be developed to promote child and parent adjustment and support the mental health of children. (Wong et al 2008)

There needs to be family directed approaches to address each family members needs to recognise and help with the symptoms of post traumatic stress disorder (Stuber et al 2006). It is important for health professionals to understand that they can influence the emotional state of parents not just because of a nursing or medical intervention but because of the way in which they communicate and manage care. (George et al 2006)

4.4 Discussion

This descriptive review was guided by three key questions with evidence being sourced from the UK only to explore the evidence base for definitions, concepts and epidemiology. International literature was also sourced to identify psychological problems. It would be interesting to identify what epidemiological evidence base is available for children's palliative care in other countries and whether the same difficulties have been encountered. It would also be useful to identify whether a solution to the complexity of counting children with palliative care needs and having a centralised system for managing this data has been established. Meeting the physical, emotional, social and spiritual needs of the child and family is part of the core focus of providing good palliative care however there is some evidence that providers of children's palliative care could benefit from further education and training so that they can more effectively identify and manage the psychological problems that may occur with the child and family.

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4.5 Conclusion

The findings suggest that the key concepts of children's palliative care tend to be reflected in the majority of the literature pertaining to children, with some giving greater emphasis on the explicit principles of children's palliative care than others. The ACT (2009) definition is widely recognised and used within policy documents within England and Wales.

The literature demonstrates that establishing the numbers of children requiring palliative care in the UK has not been easy and estimated figures using a formula derived from a number of different data collection methods provides us with some baseline for service planning. Whilst data on children is collected via a number of different sources and sectors there is no one single route or provider that can universally provide this information. Making comparisons between data found in one region compared to another is also complicated as the census points for the collection of the data may be different. There is evidence that some actual locality based data is available for specific areas, and also national data is available for specific disease groups i.e. Cancer. Children, young people and their families requiring palliative care have a broad range of characteristics and needs and require a tapestry of services which can flex and respond in order to meet these. There is evidence that psychological problems are a significant feature in the lives of children and families with life limiting and life threatening conditions with a wide range of different disorders encountered. These impact on the whole family and are not confined to the sick child. These findings highlight the importance for all professionals who are involved in caring and supporting children and their families with palliative care needs to have education and training so that they can be alert to, recognise and proactively respond in

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managing psychological problems and be aware of the impact of their own practice on the psychology of the child and the family.

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