BMJ Open  Cancer in deceased adults with intellectual disabilities: English population-based study using linked data from three sources

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ABSTRACT

Objective To improve our understanding of cancer in adults with intellectual disabilities.

Design Population-based study using linked data about deceased adults from the Learning (Intellectual) Disabilities Mortality Review (LeDeR) programme, the national cancer registry and NHS Digital.

Setting England.

Participants 1096 adults with intellectual disabilities identified by the LeDeR programme who died between 1 January 2017 and 31 December 2019.

Outcome measure Any form of cancer listed as a long-term health condition by a LeDeR reviewer or 10th edition of the International Classification of Diseases codes C00–D49 included on Parts I or II of the Medical Certificate of Cause of Death.

Results In decedents with intellectual disabilities and cancer, more than a third (35%; n=162) had cancer diagnosed via emergency presentations. Almost half (45%; n=228) of cancers were at stage IV when diagnosed. More than a third (36%; n=309) of underlying causes of deaths were of cancers of the digestive system; almost half of these (48%; n=147) were cancer of the colon, rectum or anus. Of those who died with colorectal cancer, 43% were below the age threshold for colorectal screening.

Conclusions In decedents with intellectual disabilities, symptoms suggestive of cancer had tended to be identified most frequently as an emergency and at a late stage. There is a need for greater awareness of symptoms of cancer in this population, a lower threshold for referral by General Practitioners (GPs), accelerated access to diagnosis and treatment and consideration paid to lowering the age for colorectal screening.

INTRODUCTION

We have little contemporary data about the experiences of adults with intellectual disabilities who have been diagnosed with cancer. In general, mortality studies of people with intellectual disabilities indicate a lower proportion of cancer-related deaths than in the general population, possibly due to the lower life expectancy of people with intellectual disabilities and cancer being predominantly a disease of older age. Nevertheless, cancer is one of the five most frequently recorded causes of death in studies of adults, or adults and children, with intellectual disabilities and is therefore deserving of attention in any considerations about reducing premature mortality in this population.

Within the population of people with intellectual disabilities, the risk of dying from cancer is not uniform. Females with intellectual disabilities may have increased risk of death from cancer compared with males, although this mainly appears to be a feature of younger age groups. There are conflicting findings as to whether overall cancer incidence varies by level of intellectual disability. Landes et al. reported rates of death from cancer to be higher among adults with mild or moderate intellectual disabilities than in adults with severe or profound and multiple intellectual disability or those with unspecified intellectual disability. Patja et al. found...
there to be no difference in overall cancer incidence by level of intellectual disability.

Conclusions have been drawn about the need to improve preventative measures such as cancer screening\textsuperscript{9,10} in people with intellectual disabilities, but there has been little published evidence about the diagnosis, treatment and quality of care provided for people with intellectual disabilities with cancer. In a small study of 11 women with intellectual disabilities who had breast cancer, Satgé \textit{et al},\textsuperscript{11} observed that cancers were found at a more advanced stage than in the general population. More recently, a population-based cohort study in the Netherlands concluded that cancer may be underdiagnosed and/or undertreated in people with intellectual disability.\textsuperscript{12} A scoping review of cancer treatment and survivorship among people with intellectual and developmental disabilities in the USA reported that urgent action was needed to improve collaboration among healthcare providers.\textsuperscript{13}

This paper aims to improve our understanding of cancer in adults with intellectual disabilities who have died and whose deaths were reported to the English Learning (Intellectual) Disabilities Mortality Review (LeDeR) programme. It addresses three research questions:

1. What are the types of cancer diagnosed in adults with intellectual disabilities who have died?
2. How and at what stage, was cancer diagnosed in adults with intellectual disabilities who have died?
3. What is the underlying cause of death in adults with intellectual disabilities known to have had cancer?

\section*{METHODS}

\subsection*{Study design and data}

Data about deceased adults with intellectual disabilities were extracted from the LeDeR programme reviews of deaths. The national learning from deaths policy\textsuperscript{14} requires that all deaths of people with intellectual disabilities (aged 4 years and older) in England should be reported to the LeDeR programme and reviewed using LeDeR programme methodology. The review process is described by (Heslop et al., 2020).\textsuperscript{15}

Data about the 10th edition of the International Classification of Diseases (ICD-10) codes for causes of death of people with intellectual disabilities notified to the LeDeR programme were obtained from NHS Digital. Data linkage was conducted by NHS Digital and it was based on the key identifiers of NHS number, date of birth and date of death.

For adults with intellectual disabilities for whom a LeDeR review of their death had been completed and who were known to have had cancer, data were linked to that held in the national cancer registry in England, which holds information about cancer and tumour diseases. Data linkage was conducted by the National Cancer Registration and Analysis Service and was based on the key identifiers of NHS number and date of birth. Those included were adults with intellectual disabilities who died in 2017, 2018 or 2019, for whom a LeDeR review had been completed and for whom the ICD-10 codes for cause of death had been supplied by NHS Digital. All had cancer listed as a long-term health condition by the reviewer of their death or had cancer included in either Part I or Part II of the Medical Certificate of Cause of Death (MCCD).

Data about decedents in the general population of England were drawn from data already published by the Office for National Statistics in the population data series about deaths and Public Health England cancer mortality data. Comparative data about other aspects covered in this paper were not available for decedents in England.

The ICD-10 was used to classify diagnoses and causes of death. This is divided into 21 chapters; Chapter 2 relates to neoplasms (cancer) with the codes C00–D49.

The ‘stages’ of cancer, which describe the size of the cancer and its spread, described by Cancer Research UK\textsuperscript{16} are as follows:

\begin{itemize}
  \item Stage I—the cancer is small and has not spread.
  \item Stage II—the cancer has grown but has not spread.
  \item Stage III—the cancer is larger and may have spread to the surrounding tissues and/or the lymph nodes.
  \item Stage IV—the cancer has spread from where it started to at least one other body organ, known as ‘secondary’ or ‘metastatic’ cancer.
\end{itemize}

Stage 0 refers to ‘carcinoma in situ’, ‘precancerous changes’ or ‘non-invasive cancer’. Many of these will never develop into cancer and for this reason we have not included them in this study.

We used the WHO’s definition of the underlying cause of death: the disease or injury which initiated the train of events leading directly to death.\textsuperscript{17} Although we are aware that the underlying cause of death in some people with intellectual disabilities may be inaccurately reported,\textsuperscript{18–20} such evidence does not specifically implicate the reporting of deaths from cancer, so we did not amend any cause of death reports.

We used the European harmonised definition of avoidable mortality and list of causes of death that has been adopted by the UK Office for National Statistics.\textsuperscript{21}

\begin{itemize}
  \item Preventable mortality: Causes of death that can be mainly avoided through effective public health and primary prevention interventions (ie, before the onset of diseases/injuries, to reduce incidence).
  \item Treatable mortality: Causes of death that can be mainly avoided through timely and effective health care interventions, including secondary prevention and treatment (ie, after the onset of diseases, to reduce case-fatality).
  \item Avoidable mortality: Avoidable causes of deaths are all those defined as preventable or treatable.
\end{itemize}

\section*{Outcome}

The outcome of interest was any form of cancer listed as a long-term health condition by a LeDeR reviewer or ICD-10 codes C00–D49 included on Part I or Part II of the
MCCD, of deaths occurring during the period 1 January 2017 to 31 December 2019.

**Exposure**
The definition of intellectual disabilities used was the presence of a significantly reduced ability to understand new or complex information, to learn new skills (impaired intelligence), with a reduced ability to cope independently (impaired social functioning) which started before adulthood, with a lasting effect on development.22

**Covariates**
The covariates and their sources were:
- LeDeR review of death: demographic information—age; gender; ethnicity; level of intellectual disabilities; geographic area of residence and usual type of accommodation.
- NHS Digital: ICD-10 codes for causes of death recorded on the MCCD.
- National cancer registry: relevant timescales (eg, patient age at diagnosis); how the cancer was diagnosed and the site and stage of the cancer when it was diagnosed.

**Patient and public involvement**
Family members of people with intellectual disabilities were involved in the conceptualisation of the study and in discussing the findings.

**Data analysis**
Data analysis was undertaken by analysts at NHS South Central and West Commissioning Support unit. The analysts worked with the LeDeR team at University of Bristol to ensure a full understanding of the data and to agree how it was reported. Statistical analyses were carried out using the R programming language. Initial \( \chi^2 \) analyses were carried out on frequency tables as a whole. If there was a significant effect, pairwise \( \chi^2 \) analyses were performed to determine which particular variables had significantly different proportions. For brevity we present only significant pairwise analyses results.

All numbers fewer than 10 have been suppressed to protect confidentiality.

Where appropriate, data about underlying causes of death have been compared with published general population data.

We present the findings for three distinct but interrelated groups of adults with intellectual disabilities:
- Group 1: Adults with intellectual disabilities known from LeDeR data to have died with cancer (n=1096).
- Group 2: Adults with intellectual disabilities known from LeDeR data to have died with cancer and for whom linked data were available from the national cancer registry (n=771).
- Group 3: Adults with intellectual disabilities known from LeDeR data to have died with cancer and for whom official cause of death coding from NHS Digital indicated cancer as their underlying cause of death (n=852).

**RESULTS**
Demographic data about adults with intellectual disabilities included in each group of the study are presented in table 1. Overall, there was little difference in the demographic information relating to those in groups 1, 2 and 3, although the median age at death of those in group 3 was slightly lower than that of group 1 and group 2.

**Adults with intellectual disabilities known from LeDeR data to have died with cancer (group 1)**
There were 1096 adults with intellectual disabilities who were known from LeDeR data to have died with cancer. Demographic data for adults with intellectual disabilities in group 1 were similar to the overall adult population of people with intellectual disabilities whose deaths were reported to the LeDeR programme in 2018.

Most (70%; n=771) of those included in group 1 had linked death data available from the national cancer registry and form group 2. However, 325 (30%) adults were known by the LeDeR programme to have died with cancer, but data were not available about them in the national cancer registry.

The majority of those for whom data in the national cancer registry were not available (70%, n=229) had died in 2019, a year for which registrations in the national cancer registry had not yet been completed at the time of writing. Thus, it is likely that these deaths were late registrations rather than omissions from the registry.

Data were not available from the national cancer registry for 96 (9%) adults who died in 2017 or 2018 and who were known by the LeDeR programme to have had cancer. These people tended to be older (16% aged 80 years or over, compared with 6% of those in the registry) and more had severe or profound and multiple intellectual disabilities (36%) than those in the registry (19%). A quarter (25%) of those not in the cancer registry had an 'unknown' tumour type, compared with none of those included.

**Adults with intellectual disabilities known from LeDeR data to have died with cancer and for whom linked data were available from the national cancer registry (group 2)**
There were 771 adults with intellectual disabilities known from LeDeR data to have died with cancer and for whom linked data were available from the national cancer registry (Group 2).

Information about the type of cancer diagnosed was available for all 771 of those in group 2 (table 2). Most (89%, n=690) had been diagnosed with one type of cancer; 9% (n=69) with two and 2% (n=12) with three or more different types. Thus, in the 771 people, 865 cancers had been diagnosed.

In males, the most frequently recorded types of cancers were of the digestive organs (28%); skin (12%); lip, oral cavity and pharynx (11%) and male genital organs (10%). In females, the most frequently recorded types of cancer were of the breast (26%); digestive organs (23%) and female genital organs (14%).
| Table 1 | Demographic information about people with intellectual disabilities included in the study |
|---------|----------------------------------------------------------------------------------|
| Group 1: Adults with intellectual disabilities known from LeDeR data to have died with cancer (n=1096). |
| Group 2: Adults with intellectual disabilities known from LeDeR data to have died with cancer and for whom linked data were available from the national cancer registry (n=771). |
| Group 3: Adults with intellectual disabilities known from LeDeR data to have died with cancer and for whom official cause of death coding from NHS Digital indicated cancer as their underlying cause of death (n=852). |

|                | Group 1 | Group 2 | Group 3 |
|----------------|---------|---------|---------|
| **Gender**     |         |         |         |
| Males          | 581     | 416     | 457     |
| Females        | 514     | 354     | 394     |
| **Age group (years)** |         |         |         |
| 20–34          | 44      | 31      | 38      |
| 35–49          | 123     | 75      | 104     |
| 50–64          | 430     | 322     | 369     |
| 65–79          | 420     | 296     | 300     |
| 80+            | 79      | 47      | 41      |
| **Total**      | 1096    | 771     | 852     |
| **Median age of death (years)** | 63      | 63      | 61      |
| **Ethnicity**  |         |         |         |
| White British  | 1034    | 728     | 803     |
| Non-white British | 45     | 32      | 35      |
| **Total**      | 1079    | 760     | 838     |
| **Unknown/missing** | 17      | 11      | 14      |
| **Level of intellectual disability** |         |         |         |
| Mild           | 453     | 339     | 360     |
| Moderate       | 345     | 238     | 268     |
| Severe         | 183     | 112     | 140     |
| Profound/multiple | 44     | 26      | 31      |
| **Total**      | 1025    | 715     | 799     |
| **Unknown/missing** | 71      | 56      | 53      |
| **Geographic area** |         |         |         |
| Midlands       | 214     | 147     | 171     |
| North east and Yorkshire | 168     | 123     | 131     |
| South east     | 181     | 123     | 147     |
| North west     | 160     | 111     | 125     |
| East of England | 139     | 92      | 107     |
| London         | 128     | 96      | 94      |
| South west     | 106     | 79      | 77      |
| **Total**      | 1096    | 771     | 852     |
| **Accommodation** |         |         |         |
| Supported living | 258     | 163     | 198     |
| Own or family  | 207     | 137     | 175     |
| Residential home | 190     | 122     | 145     |
| Nursing home   | 109     | 67      | 89      |
| Other          | 14      | 11      | 11      |
| **Total**      | 778     | 500     | 618     |
| **Unknown/missing** | 318     | 271     | 234     |

*There is one person whose gender was not recorded.
†Numbers fewer than 10 have been suppressed.
LeDeR, Learning (Intellectual) Disabilities Mortality Review.
Of the 771 adults with intellectual disabilities in group 2, information about the route to diagnosis was available for 60% (n=462). In these 462 adults with intellectual disabilities, cancers were more likely to be diagnosed via emergency presentations than any other route: 35% of adults with intellectual disabilities had their cancer diagnosed via an emergency referral or attendance, compared with 27% diagnosed via a non-urgent referral by their GP and 25% diagnosed via an urgent referral (table 3).

Adults with intellectual disabilities who were diagnosed via an emergency route were disproportionately male (20%, compared with 15% females; $\chi^2 (5, N=462)=29.95, p=0.0042$); younger in age (51% in the 20–49 years age group, compared with 33% of those in older age groups; $\chi^2 (20, N=462)=26.65, p=0.0017$) and living in their own or the family home (36%, compared with 16% of those living in residential care setting; $\chi^2 (25, N=462)=64.68, p=0.0014$).

Sixteen adults with intellectual disabilities had their cancer identified by screening. Of these, 14 were diagnosed with breast cancer, a rate comparable with the proportion of cancers identified by screening in the general population.

### Table 2

| ICD-10 section | Section heading | Males |   | Females |   | Total* |   |
|----------------|-----------------|-------|---|---------|---|--------|---|
| C15–C26       | Malignant neoplasms of digestive organs | 131   | 28%| 92      | 23%| 224    | 26%|
| C50           | Malignant neoplasms of breast | †     |   | †       |   | 103    | 26%|
| C43–C44       | Melanoma and other malignant neoplasms of skin | 56    | 12%| 32      | 8% | 88     | 10%|
| C00–C14       | Malignant neoplasms of lip, oral cavity and pharynx | 52    | 11%| 13      | 3% | 65     | 8% |
| C30–C39       | Malignant neoplasms of respiratory and intrathoracic organs | 39    | 8% | 21      | 5% | 60     | 7% |
| C42           | Malignant neoplasms of the haematopoietic and reticuloendothelial system | 33    | 7% | 22      | 6% | 55     | 6% |
| C51–C58       | Malignant neoplasms of female genital organs | 47    | 10%| 47      | 5% | 94     | 10%|
| C60–C63       | Malignant neoplasms of male genital organs | 33    | 7% | 13      | 3% | 46     | 5% |
| C64–C68       | Malignant neoplasms of urinary tract | †     |   | †       |   | 13     | 3% |
| C76–C80       | Malignant neoplasms of ill-defined, other secondary and unspecified sites | 24    | 5% | 13      | 3% | 37     | 4% |
| C81–C96       | Malignant neoplasms of lymphoid, haematopoietic and related tissue | 19    | 4% | 15      | 4% | 34     | 4% |
| C69–C72       | Malignant neoplasms of eye, brain and other parts of central nervous system | 20    | 4% | 10      | 3% | 30     | 3% |
| C40–41; C45–49; C73–75; D00–09; D10–36; D37–48; D49 | All other cancers | 11    | 2% | †       | †  | 20     | 2% |
|               | Total number of cancers in the 771 people | 467   | 100% | 397   | 100% | 865 | 100% |

*There is one person whose gender was not recorded. They have been included in the Total column.
†Numbers fewer than 10 have been suppressed.

ICD-10, 10th edition of the International Classification of Diseases; LeDeR, Learning (Intellectual) Disabilities Mortality Review.
Of the 865 cancers diagnosed in the 771 adults with intellectual disabilities in group 2, information about the stage of the cancer when it was diagnosed was available for 58% (n=502) of the different cancers. Almost half (46%) of the cancers diagnosed were at stage IV when the cancer had already metastasised; two-thirds (66%) were at stage III or IV (table 4).

There was some variation in the stage of cancer in adults with intellectual disabilities in group 2. Those cancers diagnosed at stage IV were disproportionately in males (51%, compared with 39% in females; $\chi^2 (8, N=503)=15.92, p=0.0033$); in adults in younger age groups (52% aged 20–49 years, compared with 45% in older age groups $\chi^2 (16, N=503)=27.18, p=0.002$) and of cancer of the digestive organs (57% of cancers of the digestive system were in these sites, a significantly greater proportion than expected proportion of cancers in adults with intellectual disabilities, genital cancer and respiratory cancer were significantly lower than in males in the general population. In females with intellectual disabilities, respiratory cancer was significantly lower than in females in the general population.

Specific ICD-10 codes for underlying causes of death from cancers of the digestive system

Disaggregating the ICD-10 cancer sections was limited by the small numbers in some sections. We therefore focused on cancers of the digestive system, the most frequently reported cancer-related underlying cause of death in adults with intellectual disabilities and more frequently reported in adults with intellectual disabilities than the general population. Three types of cancer accounted for 82% of cancers of the digestive system in adults with intellectual disabilities (table 6).

| Stage | No. | %  |
|-------|-----|----|
| I     | 88  | 18%|
| II    | 78  | 16%|
| III   | 105 | 21%|
| IV    | 228 | 45%|
| Total number of cancers | 502 | 100%|
| Unknown/missing/unstageable | 363 | |

LeDeR, Learning (Intellectual) Disabilities Mortality Review.

We compared the most frequently recorded ICD-10 sections for the cancer-related underlying causes of death for adults with intellectual disabilities in group 3 with those of the general population of England using a $\chi^2$ test. There was a significant difference between the population of adults with intellectual disabilities and adults in the general population ($\chi^2 (10, N=852)=21.79, p=0.016$). Pairwise testing suggested that in males with intellectual disabilities, genital cancer and respiratory cancer were significantly lower than in males in the general population. In females with intellectual disabilities, respiratory cancer was significantly lower than in females in the general population.
Table 5 The most frequently recorded ICD-10 codes for cancer-related underlying causes of death in adults with intellectual disabilities for whom official cause of death coding from NHS Digital indicated cancer as their underlying cause of death (group 3), compared with data reported for the general population of England

| ICD-10 codes | Neoplasm of… | LeDeR data (2017–2019) (age 18+ years) | General population (2018) (age 20+ years)† |
|--------------|---------------|----------------------------------------|----------------------------------------|
|              | Male          | Female                                 | Total*                                 |
|              | No. | %  | No. | %  | No. | %  | No. | %  | No. | %  | No. | %  | No. | %  |
| C15–C26     | Digestive organs | 191 | 42% | 117 | 30% | 309 | 36% | 23297 | 32% | 15953 | 26% | 39250 | 29% |
| C76-C80     | Ill-defined, other secondary and unspecified sites | 53  | 12%  | 32   | 8%  | 85   | 10% | 4003   | 6%  | 4448  | 7%  | 8451  | 6%  |
| C30–C39     | Respiratory and intrathoracic organs | 46  | 10%  | 23   | 6%  | 69   | 8%  | 15708  | 22% | 12910 | 21% | 28618 | 21% |
| C50         | Breast § § | 67  | 17%  | 69   | 8%  | 78   | <1%      | 9562 | 15% | 9640 | 7% |
| C81–C96; C42 | Lymphoid, haematopoietic and related tissue | 37  | 8%  | 27   | 7%  | 64   | 8%  | 3799   | 5%  | 3233  | 5%  | 7032  | 5%  |
| C51–C58     | Female genital organs | 60  | 15%  | 60   | 7%  | 6818 | 11% | 6818   | 5%  | 6818  | 5%  |
| C64–C68     | Urinary tract | 32  | 7%  | 18   | 5%  | 50   | 6%  | 5492   | 8%  | 2844  | 5%  | 8336  | 6%  |
| C69–C72     | Eye, brain and other central nervous system | 24  | 5%  | 12   | 3%  | 36   | 4%  | 2200   | 3%  | 1603  | 3%  | 3803  | 3%  |
| C60–C63     | Male genital organs | 34  | 7%  | 34   | 4%  | 10243 | 14% | 10243  | 8%  |
| C43–C44     | Melanoma and other skin | 17  | 4%  | 13   | 3%  | 30   | 4%  | 1671   | 2%  | 1007  | 2%  | 2678  | 2%  |
| C00–14; C40–41; C45–49; C73–75; D00–09; D10–36; D37–48. | All other causes | 22  | 5%  | 24   | 6%  | 46   | 5%  | 5935   | 8%  | 3691  | 5%  | 9626  | 8%  |
|             | Total         | 458 | 100% | 393 | 100% | 852 | 100% | 72426 | 100% | 62069 | 100% | 134495 | 100% |

*There is one person whose gender was not recorded. They have been included in the Total column.
†Data extracted from https://www.gov.uk/government/statistics/cancer-registration-statistics-cancer-mortality-in-england-2018.
‡Numbers fewer than 10 have been suppressed.
ICD-10, 10th edition of the International Classification of Diseases; LeDeR, Learning (Intellectual) Disabilities Mortality Review.
### Table 6  The most frequently recorded ICD-10 codes for underlying cause of death of cancer of the digestive system for adults with intellectual disabilities and general population of England

| ICD-10 codes | Neoplasm of… | LeDeR data (age 18+ years) | England (2018) (age 20+ years)† |
|--------------|---------------|-----------------------------|---------------------------------|
|              | Male % of digestive cancers | Female % of digestive cancers | Total* % of digestive cancers | Male % of digestive cancers | Female % of digestive cancers | Total % of digestive cancers |
| C18–C21 Colon, rectum and anus | 86 45% | 60 51% | 147 48% | 7509 32% | 6387 37% | 13896 34% |
| C15 Oesophagus | 43 23% | 16 14% | 59 19% | 4564 20% | 1985 11% | 6549 16% |
| C25 Pancreas | 27 14% | 18 15% | 45 15% | 4163 18% | 3868 22% | 8031 20% |
| C22 Liver and intrahepatic bile ducts | 15 8% | $§$ | 23 7% | 2907 12% | 1794 11% | 4701 12% |
| C16 Stomach | 12 6% | $§$ | 17 6% | 2197 9% | 1213 7% | 3410 8% |
| C17; C23; C24; C26 All other digestive organs | ‡ ‡ | 10 4% | 18 6% | 1957 8% | 2031 12% | 3988 10% |
| C15–C26 Digestive organs | 191 100% | 117 100% | 309 100% | 23297 100% | 17278 100% | 40575 100% |

*There is one person whose gender was not recorded. They have been included in the Total column.
†Data extracted from https://www.gov.uk/government/statistics/cancer-registration-statistics-cancer-mortality-in-england-2018
‡Numbers fewer than 10 have been suppressed.
ICD-10, 10th edition of the International Classification of Diseases; LeDeR, Learning (Intellectual) Disabilities Mortality Review.
Potentially avoidable deaths from cancer

For those in group 3 who died with cancer as an underlying cause of death, we examined the proportion of cancers known to be avoidable (either preventable or treatable), using the harmonised definition of avoidable mortality and a list of causes of death considered to be avoidable.21 Comparative information for the general population of England was not available.

Overall, 19% (n=163) adults with intellectual disabilities had a type of cancer that was a preventable cause of death. The largest proportion was due to cancer of the oesophagus (n=52; 32% of all preventable causes of death from cancer) and the lung (n=44; 27% of all preventable causes of death from cancer). Overall, 23% (n=200) of adults with intellectual disabilities had a type of cancer that was a treatable underlying cause of death. The largest proportion was due to colorectal cancer (n=106; 53% of all treatable causes of death from cancer) and breast cancer (in females only) (n=57; 29% of all treatable causes of death from cancer).

DISCUSSION

Despite the first major study of cancer in people with intellectual disabilities in England being conducted in 1997, the lack of contemporary data leads to significant policy and practice gaps. This study aimed to extend our knowledge about cancer in deceased adults with intellectual disabilities in England. One of the strengths of our study is that it links data about adults with intellectual disabilities known to have had cancer prior to their death with data from the national cancer registry and official cause of death coding from NHS Digital. This provides more comprehensive information than would otherwise be available from any of the data sources alone. There is an indication, however, that registration on the cancer registry may be incomplete for older people, those with severe or profound and multiple intellectual disabilities and those with an ‘unknown’ tumour type. We did not check the validity of the MCCD in the study population, but evidence suggests that inaccurate reporting of cause of death of people with intellectual disabilities is less likely when cancer is included as a cause of death.24 The study population was unique in that all died between 2017 and 2019; it was not a cross-sectional cohort of the population, so comparison with other findings need to be made with this caveat in mind. There were limited published data available about decedents with cancer, so comparing our data about routes to diagnosis and stage of cancer with general population data was not possible. The small number of deaths in some subcategories and when compared with general population data about underlying causes of death means that conclusions based on these data should be considered tentative.

Overall, deceased adults with intellectual disabilities more frequently had cancer diagnosed via emergency presentations than any other routes, suggesting that symptoms suggestive of cancer are not always picked up and acted on in general practice for adults with intellectual disabilities. Almost half (46%) of cancers in adults with intellectual disabilities were diagnosed at stage IV when the cancer had already metastasised to other parts of the body. Cancer survival data emphasise the importance of early diagnosis and that for most cancers, survival at 1 year and 5 years is much higher if the cancer is detected at stage I than if it is detected later.25 The NHS Long Term Plan26 states the ambition that by 2028, the proportion of cancers diagnosed at stages I and II will rise to three quarters of cancer patients. With only a third (34%) of cancers in adults with intellectual disabilities identified at these stages, there is clearly much work to do to raise greater awareness of symptoms of cancer in this population, lower the threshold for referral by GPs and accelerate access to diagnosis and treatment.

Digestive system cancers were the most common type of cancer death in adults with intellectual disabilities in this study, echoing the findings of other studies.6 8 27 This may be influenced by gastrointestinal tract dysfunction, gastro-oesophageal reflux disease and chronic constipation which are common in people with intellectual disabilities.28 There is also a suggestion that a higher incidence of these cancers in adults with intellectual disabilities, compared with the general population, may be associated with genetic deletions and family history, being overweight, inactive or having poor nutrition.29 A greater awareness of this among support staff and health professionals is important so that preventative measures can be instigated such as caregivers being vigilant about bowel habits and implementing interventions to increase fibre intake and exercise in daily activities.29

Colorectal screening presents an opportunity to discover early colorectal cancer and is available to everyone over the age of 60 years in England, with the programme expanding to include 56 years olds from 2021. Although screening rates for colorectal cancer in adults with intellectual disabilities are approaching those of the general population,30 our study found that 43% of adults diagnosed with colorectal cancer were aged 18–59 years, suggesting that the age threshold for colorectal screening in people with intellectual disabilities may need reduction.

After colorectal cancer, breast cancer was the second most frequently reported treatable cause of death in our population. Although rates of breast cancer appear to be similar in women with and without intellectual disabilities, underutilisation of breast cancer screening in adults with intellectual disabilities30 may reduce the number identified. Nulliparity, being overweight and a lack of exercise are known factors that increase breast cancer risk31 and are particularly pertinent to people with intellectual disabilities. Research is sparse about the treatment options and decision-making process for women with intellectual disabilities who have breast cancer; research relating to disabled women in general suggests that they are less likely to undergo breast-conserving surgery and...
are less likely to receive neoadjuvant chemotherapy and radiotherapy than their non-disabled peers.32

Some of the factors influencing disparities in cancer outcomes will be related to the social and economic context that shapes a person’s ability to access cancer care. Mechanisms leading to poorer outcomes in people with intellectual disabilities have been identified as provider bias and ableism;33 negative attitudes,34 the invisibility of people with intellectual disabilities;35 diagnostic overshadowing in which symptoms are misinterpreted as due to a person’s behaviour, mental state or communication impairments36 and unequal access to health services.37 Preventative measures to reduce the risk of cancer, screening and health checks to identify it early, transparent decision-making processes about options available and access to diagnostic and treatment interventions that provide a chance of optimal outcomes for people with cancer are all needed. So too is a closer research gaze on the quality of care provided to people with intellectual disabilities who have or cancer in order to ensure their equitable access to services.

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Competing interests None declared.

Patient and public involvement Patients and/or the public were involved in the design, or conduct, or reporting, or dissemination plans of this research. Refer to the Methods section for further details.

Patient consent for publication Not applicable.

Ethics approval This study involves human participants who were deceased. The Learning (Intellectual) Disabilities Mortality Review (LeDeR) programme had Section 251 approval from the national Health Research Authority’s Confidentiality Advisory Group (CAG), on behalf of the secretary of state, allowing the programme team to handle identifiable data without consent in order to conduct a review of a death and to link it to NHS Digital cause of death data. An amendment to the agreement (CAG Ref. 20CAG067) enabled the LeDeR programme to link identifiers with national cancer registry data.

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