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# 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.<sup>1</sup> In general, mortality studies of people with intellectual disabilities indicate a lower proportion of cancer-related deaths than in the general population,<sup>2 3</sup> possibly due to the lower life expectancy of people with intellectual disabilities,<sup>4 5</sup> and cancer being predominantly a

## Strengths and limitations of the study

- This study is the first to link data about adults with intellectual disabilities known to have had cancer prior to their death with that from the national cancer registry and official cause of death coding from NHS Digital.
- There is an indication 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.
- There were limited published data available about decedents with cancer, so comparing our data to general population data was not always 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.

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<sup>2 6</sup> 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,<sup>2 7</sup> although this mainly appears to be a feature of younger age groups.<sup>6</sup> There are conflicting findings as to whether overall cancer incidence varies by level of intellectual disability. Landes *et al*,<sup>2</sup> 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*,<sup>8</sup> 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<sup>9 10</sup> 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 *et al.*,<sup>11</sup> 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.<sup>12</sup> 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.<sup>13</sup>

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?

## METHODS

### 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<sup>14</sup> 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).<sup>15</sup>

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<sup>16</sup> are as follows:

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

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.<sup>17</sup> Although we are aware that the underlying cause of death in some people with intellectual disabilities may be inaccurately reported,<sup>18–20</sup> 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.<sup>21</sup>

- ▶ 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).
- ▶ 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).
- ▶ Avoidable mortality: Avoidable causes of deaths are all those defined as preventable or treatable.

### 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.<sup>22</sup>

### 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 inter-related 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.<sup>23</sup>

Most (70%; n=771) of those included in group 1 had linked 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	
	No.	%	No.	%	No.	%
<b>Gender*</b>						
Males	581	53%	416	54%	457	54%
Females	514	47%	354	46%	394	46%
<b>Age group (years)</b>						
20–34	44	4%	31	4%	38	4%
35–49	123	11%	75	10%	104	12%
50–64	430	39%	322	42%	369	43%
65–79	420	38%	296	38%	300	35%
80+	79	7%	47	6%	41	5%
Total	1096	100%	771	100%	852	100%
Median age of death (years)	63		63		61	
SD	13.33		13.34		12.78	
<b>Ethnicity</b>						
White British	1034	96%	728	96%	803	96%
Non-white British	45	4%	32	4%	35	4%
Total	1079	100%	760	100%	838	100%
Unknown/missing	17		11		14	
<b>Level of intellectual disability</b>						
Mild	453	44%	339	47%	360	45%
Moderate	345	34%	238	33%	268	34%
Severe	183	18%	112	16%	140	18%
Profound/multiple	44	4%	26	4%	31	4%
Total	1025	100%	715	100%	799	100%
Unknown/missing	71		56		53	
<b>Geographic area</b>						
Midlands	214	20%	147	19%	171	20%
North east and Yorkshire	168	15%	123	16%	131	15%
South east	181	17%	123	16%	147	17%
North west	160	15%	111	14%	125	15%
East of England	139	13%	92	12%	107	13%
London	128	12%	96	12%	94	11%
South west	106	10%	79	10%	77	9%
Total	1096	100%	771	100%	852	100%
<b>Accommodation</b>						
Supported living	258	33%	163	32%	198	32%
Own or family	207	27%	137	27%	175	28%
Residential home	190	24%	122	24%	145	23%
Nursing home	109	14%	67	13%	89	14%
Other	14	2%	11	2%	11	2%
Total	778	100%	500	100%	618	100%
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.

**Table 2** The most frequently reported cancers by ICD-10 chapter sections in 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) (n=771)

ICD-10 section	Section heading	Males		Females		Total*	
		No.	%	No.	%	No.	%
C15–C26	Malignant neoplasms of digestive organs	131	28%	92	23%	224	26%
C50	Malignant neoplasms of breast	†	†	103	26%	105	12%
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			54	14%	54	6%
C60–C63	Malignant neoplasms of male genital organs	47	10%			47	5%
C64–C68	Malignant neoplasms of urinary tract	33	7%	13	3%	46	5%
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 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 3** Route to diagnosis for adults with intellectual disabilities known from LeDeR data to have died with cancer and for whom linked data about the route to their diagnosis were available from the national cancer registry

	LeDeR data (2017–2019)	
	No.	%
Emergency presentation	162	35%
General Practitioner non-urgent referral	123	27%
Urgent referral ('2-week wait')	116	25%
Other (outpatient attendance/elective inpatient/death certificate only)	45	10%
Screening	16	3%
Total	462	100%
Unknown/missing	309	

GP, General Practitioner; LeDeR, Learning (Intellectual) Disabilities Mortality Review.

**Table 4** Stage of cancer at diagnosis for adults with intellectual disabilities known from LeDeR data to have died with cancer and for whom linked data about the route to their diagnosis were available from the national cancer registry

	LeDeR data (2017–2019)	
	No.	%
Stage I	88	18%
Stage II	78	16%
Stage III	105	21%
Stage IV	228	45%
Total number of cancers	502	100%
Unknown/missing/unstageable	363	

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 diagnosed at stage IV;  $\chi^2$  (56, N=503)=265.9, p=0.0007).

#### 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 (group 3)

##### ICD-10 chapters of underlying causes of death

Over three quarters (n=852; 78%) of the 1096 adults with intellectual disabilities in group 1 had cancer recorded as their underlying cause of death in the official cause of death coding received from NHS Digital. Small proportions had respiratory disorders (7%; n=73), circulatory disorders (4%; n=42) or other disorders (11%; n=124) recorded as their underlying cause of death.

##### ICD-10 sections of cancer-related underlying causes of death

The most frequently recorded ICD-10 section for the cancer-related underlying cause of deaths in the 852 adults in group 3 was cancer of the digestive organs (C15–26), reported for over a third (36%) of deaths (table 5). The corresponding proportion in decedents in the general population of England was 29%. The second most frequently recorded in adults with intellectual disabilities in group 3 was of

ill-defined, secondary and unspecified sites (10%), more than the corresponding proportion in the general population of England (6%), and possibly reflecting the greater than expected proportion of cancers in adults with intellectual disabilities that were diagnosed at emergency presentation and in the later stages. The third most frequently recorded in adults with intellectual disabilities in group 3 was of cancer of the respiratory and intrathoracic organs (8%), although this was substantially less than in the general population of England (21%).

There was some variation within the adults with intellectual disabilities in group 3 according to the underlying cause of death. Cancers of the digestive system were more frequently reported in males than females (42% males; 30% females;  $\chi^2$  (34, N=852)=209.63, p=0.0002) and cancers of the male genital organs were not reported in any males from minority ethnic groups ( $\chi^2$  (34, N=852)=60.262, p=0.0011).

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.

#### 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).

The most frequently recorded cancer of the digestive system was cancer of the colon, rectum and anus—almost half (48%) (n=147) of all cancers of the digestive system were in these sites, a significantly greater proportion than the 34% in the general population ( $\chi^2$  (5, N=309)=21.52, p=0.00064). The second most frequently recorded cancer of the digestive system was cancer of the oesophagus (19% (n=59) of all cancers of the digestive system), slightly more, but not significantly so, than the 16% in the general population. The third most frequently recorded was cancer of the pancreas (15% (n=45) of all cancers of the digestive system), slightly less, but not significantly so, than the proportion in the general population (20%).

**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	LeDeR data (2017–2019) (age 18+ years)						General population (2018) (age 20+ years)†					
	Male			Female			Male			Female		
	No.	%	Total*	No.	%	Total*	No.	%	Total	No.	%	Total
C15–C26	191	42%	309	117	30%	309	23297	36%	32%	15953	26%	39250
C76–C80	53	12%	85	32	8%	85	4003	10%	6%	4448	7%	8451
C30–C39	46	10%	69	23	6%	69	15708	8%	22%	12910	21%	28618
C50	§	§	69	67	17%	69	78	8%	<1%	9562	15%	9640
C81–C96; C42	37	8%	64	27	7%	64	3799	8%	5%	3233	5%	7032
C51–C58			60	60	15%	60		7%		6818	11%	6818
C64–C68	32	7%	50	18	5%	50	5492	6%	8%	2844	5%	8336
C69–C72	24	5%	36	12	3%	36	2200	4%	3%	1603	3%	3803
C60–C63	34	7%	34			34	10243	4%	14%			10243
C43–C44	17	4%	30	13	3%	30	1671	4%	2%	1007	2%	2678
C00–14; C40–41; C45–49; C73–75; D00–09; D10–36; D37–48.	22	5%	46	24	6%	46	5935	5%	8%	3691	5%	9626
Total	458	100%	852	393	100%	852	72426	100%	100%	62069	100%	134495

\*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	LeDeR data (age 18+ years)						England (2018) (age 20+ years)†											
	Male			Female			Total*			Male			Female			Total		
	No.	% of digestive cancers	% of digestive cancers	No.	% of digestive cancers	% of digestive cancers	No.	% of digestive cancers	% of digestive cancers	No.	% of digestive cancers	% of digestive cancers	No.	% of digestive cancers	% of digestive cancers	No.	% of digestive cancers	% of digestive cancers
C18–C21 Colon, rectum and anus	86	45%	51%	60	48%	48%	147	48%	7509	32%	32%	6387	37%	37%	13896	34%	34%	34%
C15 Oesophagus	43	23%	14%	16	19%	19%	59	19%	4564	20%	20%	1985	11%	11%	6549	16%	16%	16%
C25 Pancreas	27	14%	15%	18	15%	15%	45	15%	4163	18%	18%	3868	22%	22%	8031	20%	20%	20%
C22 Liver and intrahepatic bile ducts	15	8%	§	§	7%	7%	23	7%	2907	12%	12%	1794	11%	11%	4701	12%	12%	12%
C16 Stomach	12	6%	§	§	6%	6%	17	6%	2197	9%	9%	1213	7%	7%	3410	8%	8%	8%
C17; C23; C24; C26 All other digestive organs	‡	‡	4%	10	6%	6%	18	6%	1957	8%	8%	2031	12%	12%	3988	10%	10%	10%
C15–C26 Digestive organs	191	100%	100%	117	100%	100%	309	100%	23297	100%	100%	17278	100%	100%	40575	100%	100%	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.<sup>21</sup> 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.<sup>24</sup> 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.<sup>25</sup> The NHS Long Term Plan<sup>26</sup> 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.<sup>6 8 27</sup> This may be influenced by gastrointestinal tract dysfunction, gastro-oesophageal reflux disease and chronic constipation which are common in people with intellectual disabilities.<sup>28</sup> 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.<sup>29</sup> 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.<sup>29</sup>

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,<sup>30</sup> 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 disabilities<sup>30</sup> may reduce the number identified. Nulliparity, being overweight and a lack of exercise are known factors that increase breast cancer risk<sup>31</sup> 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.<sup>32</sup>

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;<sup>33</sup> negative attitudes;<sup>34</sup> the invisibility of people with intellectual disabilities;<sup>35</sup> diagnostic overshadowing in which symptoms are misinterpreted as due to a person's behaviour, mental state or communication impairments<sup>36</sup> and unequal access to health services.<sup>37</sup> 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 cancer in order to ensure their equitable access to services.

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**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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## REFERENCES

- Satgé D, Axmon A, Trétarre B, et al. Cancer diagnoses among older people with intellectual disability compared with the general population: a national register study. *J Intellect Disabil Res* 2020;64:579–88.
- Landes SD, Stevens JD, Turk MA. Cause of death in adults with intellectual disability in the United States. *J Intellect Disabil Res* 2021;65:47–59.
- Cooper S-A, Allan L, Greenlaw N, et al. Rates, causes, place and predictors of mortality in adults with intellectual disabilities with and without Down syndrome: cohort study with record linkage. *BMJ Open* 2020;10:e036465.
- Hosking FJ, Carey IM, Shah SM, et al. Mortality among adults with intellectual disability in England: comparisons with the general population. *Am J Public Health* 2016;106:1483–90.
- Heslop P, Blair PS, Fleming P, et al. The Confidential inquiry into premature deaths of people with intellectual disabilities in the UK: a population-based study. *Lancet* 2014;383:889–95.
- Glover G, Williams R, Heslop P, et al. Mortality in people with intellectual disabilities in England. *J Intellect Disabil Res* 2017;61:62–74.
- Arvio M, Salokivi T, Tiitinen A, et al. Mortality in individuals with intellectual disabilities in Finland. *Brain Behav* 2016;6:e00431.
- Patja K, Eero P, Iivanainen M. Cancer incidence among people with intellectual disability. *J Intellect Disabil Res* 2001;45:300–7.
- Merten JW, Pomeranz JL, King JL, et al. Barriers to cancer screening for people with disabilities: a literature review. *Disabil Health J* 2015;8:9–16.
- Osborn DPJ, Horsfall L, Hassiotis A, et al. Access to cancer screening in people with learning disabilities in the UK: cohort study in the health improvement network, a primary care research database. *PLoS One* 2012;7:e43841.
- Satgé D, Sauleau E-A, Jacot W, et al. Age and stage at diagnosis: a hospital series of 11 women with intellectual disability and breast carcinoma. *BMC Cancer* 2014;14:150.
- Cuyper M, Tobi H, Huijsmans CAA, et al. Disparities in cancer-related healthcare among people with intellectual disabilities: a population-based cohort study with health insurance claims data. *Cancer Med* 2020;9:6888–95.
- Samtani G, Bassford TL, Williamson HJ, et al. Are researchers addressing cancer treatment and survivorship among people with intellectual and developmental disabilities in the U.S.? A scoping review. *Intellect Dev Disabil* 2021;59:141–54.
- National Quality Board. National guidance on learning from deaths, 2017. Available: <https://www.england.nhs.uk/wp-content/uploads/2017/03/nqb-national-guidance-learning-from-deaths.pdf>
- Heslop P, Byrne V, Calkin R, et al. Establishing a national mortality review programme for people with intellectual disabilities: the experience in England. *J Intellect Disabil* 2020;November 2020:1744629520970365.
- Cancer research UK (not dated) stages of cancer. Available: <https://www.cancerresearchuk.org/about-cancer/what-is-cancer/stages-of-cancer>
- WHO. International statistical classification of diseases and related problems. In: *10th revision instruction manual*. Vol 2. 5 edn, 2016. [https://icd.who.int/browse10/Content/statchtml/ICD10Volume2\\_en\\_2016.pdf](https://icd.who.int/browse10/Content/statchtml/ICD10Volume2_en_2016.pdf)
- Walker AR, Trollor JN, Reppermund S, et al. Reviewing causes of death of individuals with intellectual disability in New South Wales, Australia: a record-linkage study. *J Intellect Disabil Res* 2021;65:998–1009.
- Landes SD, Turk MA, Lauer E. Recommendations for accurately reporting intellectual and developmental disabilities on death certificates. *Am J Prev Med* 2020;59:892–5.
- Landes SD, Stevens JD, Turk MA. Obscuring effect of coding developmental disability as the underlying cause of death on mortality trends for adults with developmental disability: a cross-sectional study using us mortality data from 2012 to 2016. *BMJ Open* 2019;9:e026614.
- ONS. Consultation response: review of avoidable mortality definition, 2019. Available: <https://www.ons.gov.uk/peoplepopulationandcommunity/healthandsocialcare/causesofdeath/bulletins/avoidablemortalityinenglandandwales/latest>
- Department of Health. Valuing people. A new strategy for learning disability for the 21st century., 2001. Available: [https://assets.publishing.service.gov.uk/government/uploads/system/uploads/attachment\\_data/file/250877/5086.pdf](https://assets.publishing.service.gov.uk/government/uploads/system/uploads/attachment_data/file/250877/5086.pdf)
- Heslop P, Byrne V, Calkin R, Pollard J, Sullivan B, in conjunction with the LeDeR team at the University of Bristol.. Learning disabilities mortality review (LeDeR) programme annual report 2020. Bristol University of Bristol; 2021. <https://leder.nhs.uk/resources/annual-reports>

- 24 Landes SD, Turk MA, Bisesti E. Uncertainty and the reporting of intellectual disability on death certificates: a cross-sectional study of US mortality data from 2005 to 2017. *BMJ Open* 2021;11:e045360.
- 25 Hawkes N. Cancer survival data emphasise importance of early diagnosis. *BMJ* 2019;364:l408.
- 26 NHS. The NHS long term plan, 2019. Available: <https://www.longtermplan.nhs.uk/wp-content/uploads/2019/08/nhs-long-term-plan-version-1.2.pdf>
- 27 Tyrer F, McGrother C. Cause-Specific mortality and death certificate reporting in adults with moderate to profound intellectual disability. *J Intellect Disabil Res* 2009;53:898–904.
- 28 Davis RW. Digestive system diseases. In: *Intellectual disability and ill-health: a review of the evidence*. Cambridge: Cambridge University Press, 2010.
- 29 Willis D, Samalin E, Satgé D. Colorectal cancer in people with intellectual disabilities. *Oncology* 2018;95:323–36.
- 30 NHS Digital. Health and care of people with learning disabilities experimental statistics 2019 to 2020, 2021. Available: <https://app.powerbi.com/view?r=eyJrljoiNTYyNDM4MGYtZDRmYi00NTAxLTkzY2Q0MjcwZTY2YTQ0MzNkIiwidCI6IjUwZjYwNzFmLWJiZmUtNDAXYS04ODAzLTY3Mzc0OGU2MjllMlslmMiOjh9>
- 31 Momenimovahed Z, Salehiniya H. Epidemiological characteristics of and risk factors for breast cancer in the world. *Breast Cancer* 2019;11:151–64.
- 32 McCarthy EP, Ngo LH, Roetzheim RG, *et al*. Disparities in breast cancer treatment and survival for women with disabilities. *Ann Intern Med* 2006;145:637–45.
- 33 Stirling M, Linton J, Ouellette-Kuntz H, *et al*. Scoping review protocol documenting cancer outcomes and inequalities for adults living with intellectual and/or developmental disabilities. *BMJ Open* 2019;9:e032772.
- 34 Hemm C, Dagnan D, Meyer TD. Identifying training needs for mainstream healthcare professionals, to prepare them for working with individuals with intellectual disabilities: a systematic review. *J Appl Res Intellect Disabil* 2015;28:98–110.
- 35 Hafeez S, Singhera M, Huddart R. Exploration of the treatment challenges in men with intellectual difficulties and testicular cancer as seen in Down syndrome: single centre experience. *BMC Med* 2015;13:152.
- 36 Satgé D, Kempf E, Dubois J-B, *et al*. Challenges in diagnosis and treatment of lung cancer in people with intellectual disabilities: current state of knowledge. *Lung Cancer Int* 2016;2016:1–7.
- 37 Doherty AJ, Atherton H, Boland P, *et al*. Barriers and facilitators to primary health care for people with intellectual disabilities and/or autism: an integrative review. *BJGP Open* 2020;4:bjgpopen20X101030.