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ORIGINAL ARTICLE

Breast (female), colorectal, and lung cancer survival in people with intellectual or developmental disabilities: A population-based retrospective cohort study

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Abstract

Objectives Cancer is a leading cause of death among people living with intellectual or developmental disabilities (IDD). There is little empirical evidence documenting survival or comparing outcomes to those without IDD. This study investigated the association between IDD and cancer survival among adults with breast (female), colorectal, or lung cancer.

Methods A population-based retrospective cohort study was conducted in Ontario, Canada, with routinely collected data. Patients with breast, colorectal, or lung cancer were included (2007−2019). IDD status before cancer was determined using an established administrative data algorithm. The outcomes of interest included death from any cause and death from cancer. Cox proportional hazards models and competing events analyses using multivariable cause-specific hazards regression were completed. Analyses were stratified by cancer type. Interactions with age, sex, and stage at diagnosis, as well as sensitivity analyses, were completed.

Results The final cohorts included 123,695 breast, 98,809 colorectal, and 116,232 lung cancer patients. Individuals with IDD experienced significantly worse survival than those without IDD. The adjusted hazard ratios of all-cause death were 2.74 (95% CI 2.41−3.12), 2.42 (95% CI 2.18−2.68), and 1.49 (95% CI 1.34−1.66) times higher for breast, colorectal, and lung cancer patients with IDD relative to those without. These findings were consistent for cancer-specific deaths. With few exceptions, worse survival for people with IDD persisted regardless of stage at diagnosis.

Conclusion People with IDD experienced worse cancer survival than those without IDD. Identifying and intervening on the factors and structures responsible for survival disparities is imperative.

Résumé

Objectifs Le cancer est l'une des principales causes de mortalité chez les personnes vivant avec des déficiences intellectuelles ou des troubles du développement (DI/TD). Il y a peu de preuves empiriques décrivant la survie de ces personnes lorsqu'elles sont atteintes d'un cancer ou comparant leurs résultats à ceux des personnes sans DI/TD. Notre étude porte sur l'association entre les DI/TD et la survie au cancer chez les adultes atteints de cancer du sein (femmes), du colorectum ou du poumon.

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Méthode Une étude de cohorte rétrospective populationnelle a été menée en Ontario, au Canada, à l'aide de données recueillies systématiquement. Nous avons inclus les patientes et les patients atteints de cancer du sein, du colorectum ou du poumon (2007−2019). Nous avons identifié la présence des DI/TD avant le cancer à l'aide d'un algorithme de traitement de données administratives reconnu. Les résultats d'intérêt étaient les décès de toutes causes et les décès dus au cancer. Nous avons appliqué des modèles des risques proportionnels de Cox et des analyses des événements concurrents en utilisant la régression multivariée des risques par cause. Nos analyses ont été stratifiées selon le type de cancer. Nous avons tenu compte des interactions avec l'âge, le sexe et le stade au diagnostic et effectué des analyses de sensibilité.

Résultats Les cohortes finales ont inclus 123 695 personnes atteintes de cancer du sein, 98 809 atteintes de cancer colorectal et 116 232 atteintes de cancer du poumon. La survie des sujets ayant des DI/TD a été significativement moins bonne que celle des sujets sans DI/TD. Les rapports de risques instantanés ajustés pour les décès de toutes causes étaient 2,74 fois (IC de 95 % 2,41−3,12), 2,42 fois (IC de 95 % 2,18−2,68) et 1,49 fois (IC de 95 % 1,34−1,66) plus élevés chez les personnes atteintes de cancer du sein, du colorectum et du poumon et ayant des DI/TD que chez les personnes sans DI/TD. Ces constatations ressortent pour tous les décès attribuables à des cancers particuliers. Avec peu d'exceptions, la survie moins bonne pour les personnes ayant des DI/TD persistait quel que soit le stade au moment du diagnostic.

Conclusion La survie au cancer était moins bonne chez les personnes ayant des DI/TD que chez celles n'ayant pas de DI/TD. Il est impératif d'identifier les facteurs et les structures responsables de ces disparités dans la survie et d'intervenir en conséquence.

Keywords Intellectual or developmental disability · Cancer survival · Population-based study · Retrospective cohort study · Administrative data · Equity

Mots-clés Déficiences intellectuelles ou développementales · survie au cancer · étude populationnelle · étude de cohorte rétrospective · données administratives · équité

Introduction

Approximately 1−3% of the population live with intellectual or developmental disabilities (IDD), which are characterized by lifelong differences in intellectual functioning and adaptive behaviour that appear during the developmental period (Maulik et al., 2011; Sullivan et al., 2018). Severity of the disability ranges from mild to severe and is related to both the underlying diagnosis and other social determinants of health (Sullivan et al., 2018). Individuals with IDD live on average 10 to 20 fewer years than the general population (Patja et al., 2000). Differences in life expectancy exist for several biomedical and psychosocial reasons, including higher rates of and worse outcomes from physical illnesses, such as cardiovascular disease, epilepsy, diabetes (Sullivan et al., 2018), and cancer (Glover et al., 2017). In Ontario, Canada, cancer is the second leading cause of death for persons with IDD (Stankiewicz et al., 2018).

People living with IDD may experience worse cancer survival as the cumulative effect of delayed cancer diagnoses and lower receipt of cancer treatment, resulting from a constellation of interacting factors related to their disability and their environment (Boonman et al., 2022; Samtani et al., 2021; Stirling et al., 2021). Delays in diagnoses may result from difficulty communicating with caregivers and or care providers about signs and symptoms of cancer, through a dependency on others to attend medical appointments including cancer screening, and from interfering or competing physical and mental diagnoses (Krahn et al., 2006; Ouellette-Kuntz, 2005; Sullivan et al., 2018). People with IDD may also experience challenges perceiving and processing new information that could result in the non-disclosure of cancer diagnoses and non-curative treatment decisions made by caregivers and health care professionals (Boonman et al., 2022). Adults with IDD disproportionately experience social risk factors associated with worse cancer survival (Krahn et al., 2006; Ouellette-Kuntz, 2005). A lack of health care providers' knowledge and internalized attitudes about disability, as well as systematic biases such as ableism, may further decrease the provision of tailored, patient-centered care, and create additional obstacles to receiving potentially curative treatment (Stirling et al., 2021).

Despite the plausible increased risk of poor cancer outcomes, data on cancer prognosis for people with IDD are scarce and there are no epidemiological studies comparing cancer survival for people with IDD with that for the cancer population without IDD (Boonman et al., 2022; Samtani et al., 2021; Stirling et al., 2021). Identifying cancer survival disparities is an important first step in supporting people with IDD effectively throughout the cancer care continuum. Such findings will inform strategies for creating action, supporting advocacy, and informing people living with IDD and their families of the increased risk among cancer patients living with IDD. Multiple leading cancer organizations globally emphasize the importance of cancer care equity yet have not focused on people living

with IDD; therefore, this research is imperative for creating changes in the care provided to cancer patients living with IDD who may be marginalized within the current health care systems.

The objective of this study was to test the hypothesis that overall survival and cancer-specific survival following a breast (female), colorectal, or lung cancer diagnosis are lower for people living with than for those living without IDD.

Methods

Setting and design

This was a retrospective cohort study, using population-based routinely collected data at ICES (formerly the Institute for Clinical Evaluative Sciences) in Ontario, Canada, which includes almost 13.5 million residents (40% of the Canadian population) (Statistics Canada, 2022). Canada provides universal health coverage to all citizens and permanent residents through provincial and territorial health insurance coverage. Approximately 1.8−3.6% of Canadian residents may not qualify for public health insurance (e.g., temporary workers) (Goel & Beder, 2012). ICES is an independent, non-profit research institute funded by an annual grant from the Ontario Ministry of Health and Long-Term Care. As a prescribed entity under Ontario's privacy legislation, ICES is authorized to collect and use health care data from all individuals with provincial health insurance without individual consent for the purposes of health system analysis, evaluation, and decision support. Secure access to these data is governed by policies and procedures that are approved by the Information and Privacy Commissioner of Ontario. The study received ethical clearance from the University of Manitoba Health Research Ethics Board (#H2019:253) and Queen's University Faculty of Health Sciences and Affiliated Hospitals Research Ethics Board (#EPID-691-19). This study was reported according to the STROBE guideline for cohort studies (von Elm et al., 2007).

Data sources

Multiple databases across Ontario were linked using unique encoded identifiers and analyzed at ICES (Supplementary Table S1). Data sources included the cancer and the vital statistics registry, hospitalization, physician billing, emergency department, home care, and complex continuing care records, and the Registered Persons Database (RPDB). The Ontario Cancer Registry (OCR) has captured cancer diagnoses since 1964 with a 98% completeness rate and cancer stage data since 2007 (Clarke et al., 1991; Robles et al., 1988).

Study populations

The study cohorts included adult residents aged ≥ 18 years with a malignant breast (female), colorectal, or lung cancer recorded in the OCR with a diagnosis date between 01/01/2007 and 12/31/2019. Breast (female), colorectal, and lung cancers were explored as they represent three of the four most common cancer diagnoses in Canada (Brenner et al., 2022), have reliable cancer staging data available at the population level in the OCR, and represent cancers with and without population-level screening programs, as well as those that occur within and across sexes. Cohort members were identified using ICD-O-3 codes: breast: C500-506, C508-509; colorectal: C180, C182-89, C199, C209, C260; lung: C340-343, C348-349. Individuals were excluded if they met any of the following criteria: (1) non-resident; (2) invalid death date; or (3) no health insurance coverage on index date or in the 2 years prior to index.

Determining IDD status

We applied an established administrative data algorithm from birth or the start of data availability, up until the 6 months prior to the cancer diagnosis to identify people living with IDD (Lin et al., 2013; Ouellette-Kuntz & Lynn, 2014). To be classified as meeting the definition of having an IDD, we required an individual to have ≥ 1 hospital admission, ≥ 1 complex continuing care admission, ≥ 1 emergency department visit, ≥ 1 home care visit, or ≥ 2 physician visits with an eligible diagnosis code recorded alongside a health encounter prior to the cancer diagnosis. Diagnostic codes were recorded in forms specific to the data source, including International Classification of Diseases revisions 9 and 10 (hospitals, emergency department visits, physician visits) and administrative binary yes/no variables (complex continuing care, home care). Eligible diagnoses included intellectual disabilities, fetal alcohol syndrome, autism, Down syndrome, and others; a list of codes is provided in Supplementary Tables S2-3. Individuals who did not meet the definition were classified in the reference group (without IDD). Severity of the IDD could not be categorized, nor could the specific diagnosis be captured by the IDD definition with available data. We completed sensitivity analysis for the definition of IDD whereby IDD determination continued past the cancer diagnosis until the end of the follow-up period.

Outcomes

Death from any cause and cancer-specific death were the outcomes of interest. Death from any cause was identified from the RPDB, which includes data on vital status up until

12/31/2021. Individuals were followed from their date of index cancer diagnosis until they died or 12/31/2021 (whichever came first). Cause of death was identified from the vital statistics registry and was only available for a subgroup of individuals diagnosed with cancer between 01/01/2007 and 12/31/2018. The primary definition of cancer-specific death included any cancer diagnosis code in the primary cause of death (ICD-9 140–239). As a sensitivity analysis, we modified the definition to restrict eligible ICD-9 codes specific to each primary cancer site (ICD 9 codes breast (174), colorectal (153, 154) and lung (162 excluding 162.0, 162.2)).

Covariates

Covariates were measured in the year of cancer diagnosis except for comorbidity which was measured in the 2 years prior to the cancer diagnosis. Sociodemographic information included age, sex, rurality, region, and community-level income. Rurality was examined using the Rurality Index of Ontario score (RIO) (Kralj, 2009). For region, people were categorized into one of 14 geographic regions called Local Health Integration Network (LHIN) based on the individual's postal code at index. Community-level income was estimated by linking neighbourhood level census income data with postal codes and reported in quintiles (1=lowest; 5=highest). Using the Johns Hopkins ACG® system, major and minor physical comorbidities were estimated into Aggregate Diagnosis Groups (ADGs) (Johns Hopkins University, 2014). Six major and 22 minor ADGs were described based on type, diagnosis, and the number of interventions and encounters. Cancer stage at diagnosis was classified according to the appropriate AJCC/UICC Edition and available from the OCR.

Statistical analysis

All analyses were performed separately for each of the breast, colorectal, and lung cancer cohorts. We summarized baseline characteristics stratified by IDD status and compared exposure groups using standardized differences (values ≥ 0.1 were considered significant) (Austin, 2009). Kaplan-Meier survival curves for all-cause death were plotted. Log-rank tests comparing strata by IDD status and 5-year survival rates were reported for each cancer type. The association between IDD and all-cause death was estimated with multivariable Cox proportional hazards regression. Censoring occurred at the end of the follow-up period. Hazard ratios (HR) with 95% confidence intervals (CIs) were estimated. Multivariable models were adjusted for age (continuous), sex (colorectal, and lung), rurality, geography, year of diagnosis, previous cancer, small-cell cancer (lung), and colon cancer (colorectal). Confounder selection from established risk factors for worse cancer survival was informed by the principles of effect decomposition for health equity research outlined by Jackson (2021). Under this framework, confounder selection not only should be informed by traditional causal inference definitions but also requires the consideration of whether differences in the distribution of the potential conditioning factor are unfair (Jackson, 2021). For example, measures of access to health care (receipt of cancer screening, stage at diagnosis), health and lifestyle behaviours (comorbidity), and social vulnerabilities (income) were considered contributing factors modifiable by upstream health and social programs and non-allowable for inclusion as confounders in our statistical models. The association between IDD and cancer-specific death was estimated using multivariable cause-specific Cox proportional hazards regression (Austin et al., 2016), with death from non-cancer causes as the competing event. Individuals were censored at the date of non-cancer death, or end of the follow-up period, whichever came first. Cause-specific hazard ratios and 95% CI were estimated and adjusted for the same set of confounders. Effect heterogeneity by age at diagnosis, sex, and stage at diagnosis was investigated through inclusion of interaction terms in the multivariable cause-specific models. Strata-specific estimates and p-values were estimated from the full model. Statistical analyses were completed using SAS Version 9.4. Statistical significance was set at α=0.05. The proportional hazards assumption was assessed visually in the Kaplan-Meier curves and using Schoenfeld's test (Hess, 1993). We had 80% power to detect a HR of 1.5 with a two-tailed alpha of 0.05 and assuming a 1% prevalence of IDD in the study cohort (see supporting material).

Missing data

There were no missing data on IDD status, vital status, cause of death, or included covariates in the model. Missing data on descriptive variables are presented in Table 1.

Results

The final cohorts for all-cause death included 123,695 people diagnosed with breast (female) cancer, 98,809 people diagnosed with colorectal cancer, and 116,232 people diagnosed with lung cancer in Ontario between 2007 and 2019 (Supplementary Figure S1). Within these cohorts, 486 (0.39%), 506 (0.51%), and 385 (0.33%) met the IDD definition, respectively. Median follow-up times were 7.3 years, 7.3 years, and 5.4 years for survivors, respectively. Table 1 summarizes baseline characteristics, stratified by IDD status.

Overall survival differed by IDD status across cancer cohorts (p<0.001 for all; Fig. 1a–c). Five-year survival for people with IDD was 61.5% for those with breast cancer, 34.2% for those with colorectal cancer, and 11.9% for those with lung cancer compared with 81.7%, 56.6%, and 19.7%

among those without IDD, respectively. Crude and adjusted hazards ratios are displayed in Table 2. After adjustment for confounders, those with IDD were 2.74 (95% CI 2.41−3.12), 2.42 (95% CI 2.18−2.68), and 1.49 (95% CI 1.34−1.66) times more likely to die following their breast cancer, colorectal cancer, and lung cancer diagnoses, compared with those without IDD. Statistical variation in the hazard ratios was documented over time (Supplementary Figure S2a–c).

The cancer-specific survival analysis included 112,775 patients with breast (female) cancer, 91,411 with colorectal cancer, and 107,159 with lung cancer. Of these subcohorts, 444 (0.39%) patients with breast, 461 (0.50%) with colorectal, and 355 (0.33%) with lung cancer met the definition of IDD respectively. Median follow-up times in the cancer-specific cohort were 4.6, 4.4, and 2.0 years for cancer survivors with breast (female), colorectal, and lung cancers. Cumulative incidence of cancer-specific death differed significantly by IDD status across the breast (female) and colorectal cancer cohorts but not among the lung cancer cohort ($p<0.001$ for breast and colorectal; $p=0.0550$ for lung; Supplementary Figures S3–S5). Crude and adjusted cause-specific hazard ratios are presented in Table 3. After adjustment for confounders, people living with IDD were 2.28 times more likely to die of breast cancer (95% CI 1.86−2.78), 2.57 times more likely to die of colorectal cancer (95% CI 2.26−2.92), and 1.38 times more likely to die of lung cancer (95% CI 1.21−1.57) during the study period than those without IDD. The results were robust to sensitivity analyses varying the IDD determination time frame and to restriction of the cancer-specific death definition to

Table 1 Baseline characteristics among Ontarian cancer patients, stratified by intellectual or developmental disability (IDD) status

Variable	Value	Breast cancer cohort			Colorectal cancer cohort			Lung cancer cohort		
		IDD	No IDD	SD	IDD	No IDD	SD	IDD	No IDD	SD
		N=486 *N* (%)	*N*=123,209 *N* (%)		*N*=506 *N* (%)	*N*=98,303 *N* (%)		*N*=385 *N* (%)	*N*=115,847 *N* (%)	
Age (years)	≤49	93 (19.1)	22,799 (18.5)	0.02	70 (13.8%)	7065 (7.2)	0.22	23 (6.0)	3328 (2.9)	0.15
	50–59	138 (28.4)	28,841 (23.4)	0.11	113 (22.3%)	15,250 (15.5)	0.17	69 (17.9)	14,738 (12.7)	0.14
	60–69	122 (25.1)	32,030 (26.0)	0.02	122 (24.1%)	23,704 (24.1)	0	117 (30.4)	33,030 (28.5)	0.04
	70–79	68 (14.0)	23,694 (19.2)	0.14	111 (21.9%)	27,118 (27.6)	0.13	110 (28.6)	38,985 (33.7)	0.11
	≥80	65 (13.4)	15,845 (12.9)	0.02	90 (17.8%)	25,166 (25.6)	0.19	66 (17.1)	25,766 (22.2)	0.13
Female		486 (100)	123,209 (100)	--	221 (43.7%)	44,846 (45.6)	0.04	177 (46.0)	56,679 (48.9)	0.06
Rurality[A]	Rural	58 (11.9)	11,241 (9.1)	0.09	84 (16.6%)	11,714 (11.9)	0.13	63 (16.4)	14,936 (12.9)	0.1
	Semi-urban	230 (47.3)	60,104 (48.8)	0.03	244 (48.2%)	47,740 (48.6)	0.01	203 (52.7)	57,327 (49.5)	0.06
	Urban	198 (40.7)	51,864 (42.1)	0.03	178 (35.2%)	38,849 (39.5)	0.09	119 (30.9)	43,584 (37.6)	0.14
Income quintile[B]	1	132 (27.3)	22,015 (17.9)	0.23	140 (27.7%)	19,740 (20.1)	0.18	126 (33.1)	28,104 (24.3)	0.19
	2	108 (22.3)	24,316 (19.8)	006	112 (22.2%)	20,515 (20.9)	0.03	85 (22.3)	25,791 (22.3)	0
	3	98 (20.2)	24,118 (19.6)	0.02	94 (18.6%)	19,518 (19.9)	0.03	58 (15.2)	22,534 (19.5)	0.11
	4	72 (14.9)	25,392 (20.7)	0.15	71 (14.1%)	19,175 (19.6)	0.15	68 (17.8)	20,520 (17.8)	0
	5	74 (15.3)	27,053 (22.0)	0.17	88 (17.4%)	19,040 (19.4)	0.05	44 (11.5)	18,506 (16.0)	0.13
TNM stage	0/I	169 (34.8)	53,196 (43.2)	0.17	66 (13.0%)	19,416 (19.8)	0.18	59 (15.3)	20,244 (17.5)	0.06
	II	152 (31.3)	39,449 (32.0)	0.02	87 (17.2%)	22,193 (22.6)	0.14	23 (6.0)	7529 (6.5)	0.02
	III	69 (14.2)	14,246 (11.6)	0.08	113 (22.3%)	25,063 (25.5)	0.07	59 (15.3)	20,262 (17.5)	0.06
	IV	29 (6.0)	5699 (4.6)	0.06	108 (21.3%)	16,520 (16.8)	0.12	115 (29.9)	48,345 (41.7)	0.25
	Unknown	67 (13.8)	10,619 (8.6)	0.16	132 (26.1%)	15,111 (15.4)	0.27	129 (33.5)	19,467 (16.8)	0.39
# Major ADGs[C]	0	103 (21.2)	47,190 (38.3)	0.38	69 (13.6%)	24,064 (24.5)	0.28	13 (3.4)	13,962 (12.1)	0.33
	1	148 (30.5)	39,950 (32.4)	0.04	107 (21.1%)	27,680 (28.2)	0.16	48 (12.5)	29,826 (25.7)	0.34
	2+	235 (48.4)	36,069 (29.3)	0.4	330 (65.2%)	46,559 (47.4)	0.37	324 (84.2)	72,059 (62.2)	0.51
# Minor ADGs[C]	0–2	61 (12.6)	11,523 (9.4)	0.1	16 (3.2%)	3918 (4.0)	0.04	17 (4.4)	4856 (4.2)	0.01
	3–4	85 (17.5)	26,563 (21.6)	0.1	54 (10.7%)	14,470 (14.7)	0.12	36 (9.4)	15,760 (13.6)	0.13
	5–6	92 (18.9)	32,962 (26.8)	0.19	134 (26.5%)	25,570 (26.0)	0.01	85 (22.1)	28,466 (24.6)	0.06
	≥7	248 (51.0)	52,161 (42.3)	0.17	302 (59.7%)	54,345 (55.3)	0.09	247 (64.2)	66,765 (57.6)	0.13

ADG aggregate diagnosis groups, *SD* standardized difference, *IDD* intellectual or developmental disability

[A]Missing are those most remote and least connected to health services and classified as rural

[B]Missing *n*=315 for breast cancer cohort; *n*=315 for colorectal cancer cohort; *n*=392 for lung cancer cohort

[C]Using the Johns Hopkins ACG® system

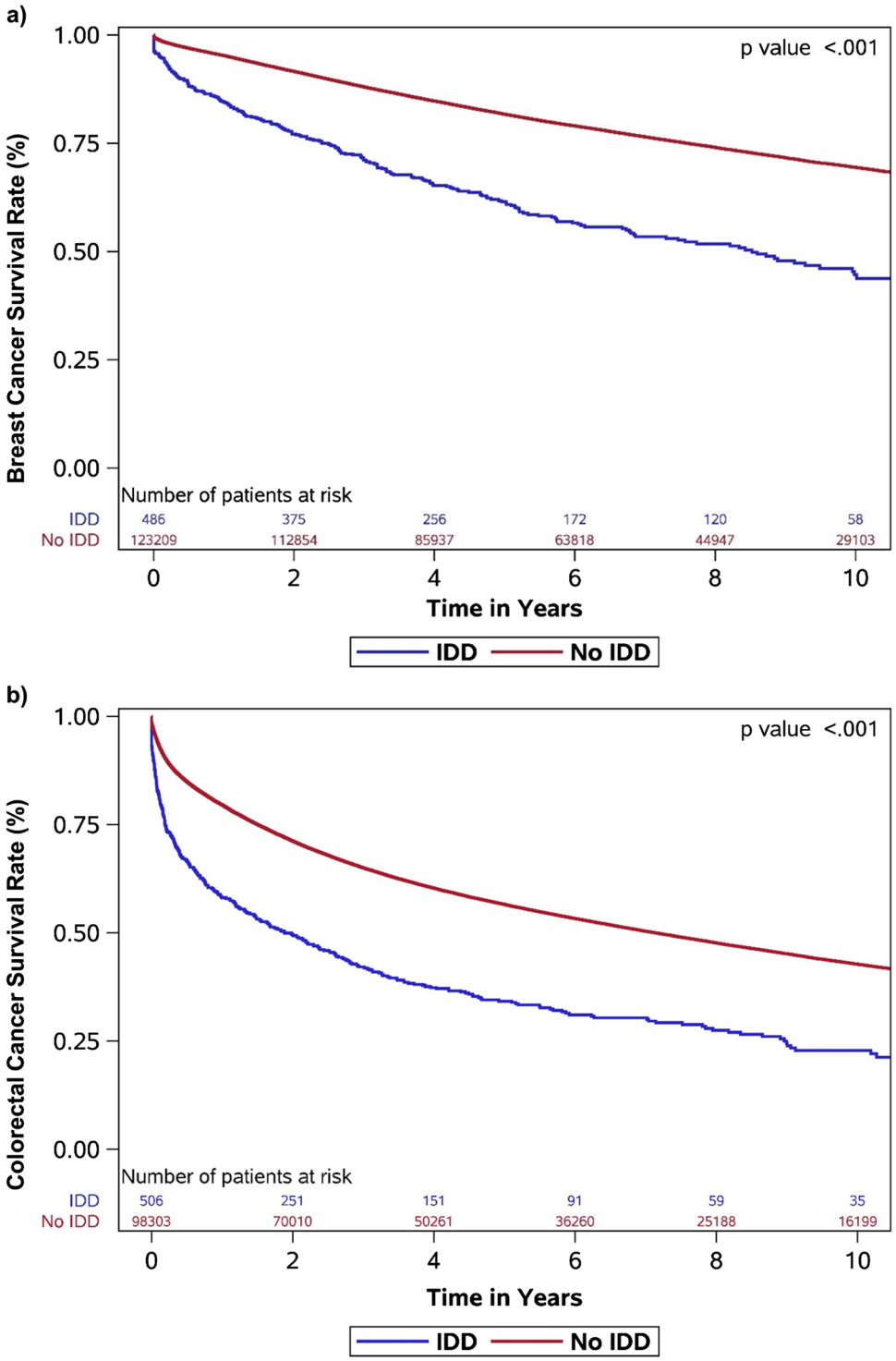

Fig. 1 **a** Kaplan-Meier plot comparing overall survival rate among breast cancer patients with and without IDD. **b** Kaplan-Meier plot comparing overall survival rate among colorectal cancer patients with and without IDD. **c** (see next page) Kaplan-Meier plot comparing overall survival rate among lung cancer patients with and without IDD

those attributed to the primary cancer site (Supplementary Table S4).

Table 4 summarizes our findings related to differences in the association between IDD status and cancer-specific survival by age, sex, and stage of cancer at diagnosis. We observed that the effect estimates were consistent across age categories (excluding ≤ 49 for lung cancer: HR= 0.89; 95% CI 0.48−1.66) and in both males and females. We identified significant effect heterogeneity in the association between IDD status and cancer-specific survival across TNM stage categories for colorectal cancer and a similar signal for breast cancer. Supplementary Table S5 describes the number of cancer deaths by stage for each cancer

Fig. 1 (continued)

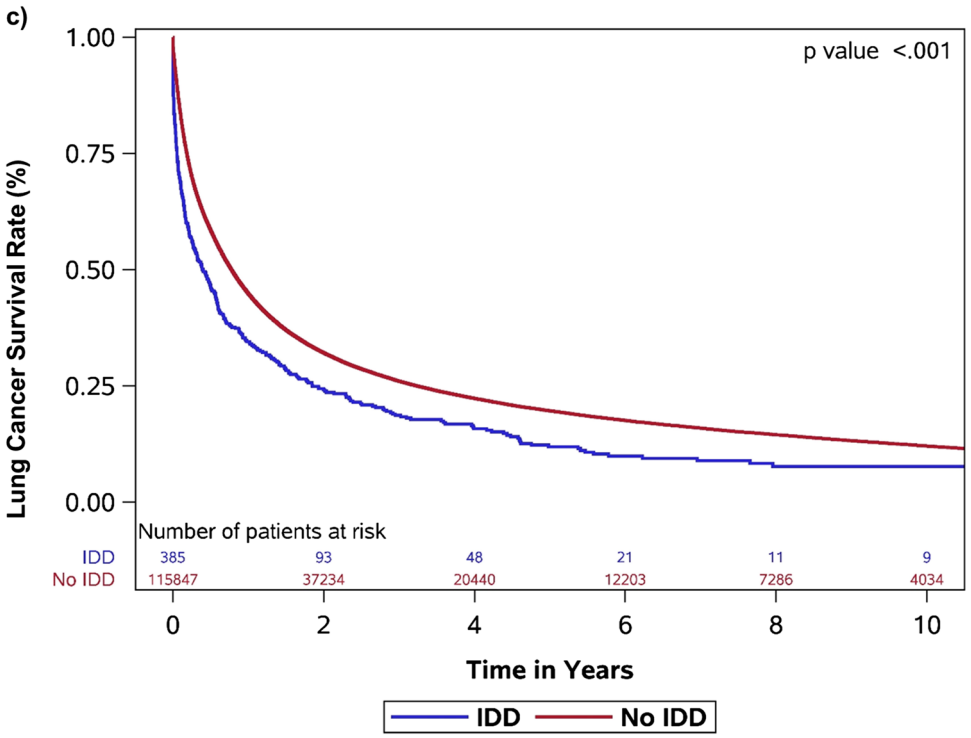

Table 2 Association between intellectual or developmental disability (IDD) status and overall survival

	N	*N* event (%)	Crude HR (95% CI)	*p*	Adjusted HR[D] (95% CI)	*p*
Breast						
IDD	486	228 (46.9)	2.37 (2.08–2.71)	<0.001	2.74 (2.41–3.12)	<0.001
No IDD	123,209	31,266 (25.4)	Reference		Reference	
Colorectal						
IDD	506	356 (70.4)	1.96 (1.77–2.18)	<0.001	2.42 (2.18–2.68)	<0.001
No IDD	98,303	51,179 (52.1)	Reference		Reference	
Lung						
IDD	385	345 (89.6)	1.37 (1.23–1.52)	<0.001	1.49 (1.34–1.66)	<0.001
No IDD	115,847	96,417 (83.2)	Reference		Reference	

HR hazard ratio, *CI* confidence interval, *IDD* intellectual or developmental disability

[D]Adjusted for age (continuous), rurality, year of diagnosis, geography, sex *(Colorectal* and *Lung*), previous cancer, small-cell cancer (*Lung*), and colon cancer (*Colorectal*)

site, by IDD status. Heterogeneity appeared to be driven by a larger magnitude of the associations for unknown stage, relative to other stages of disease. In breast cancer, there also appeared to be no effect of IDD status on cancer-specific survival in stage 0/I disease. In colorectal cancer, the effect of IDD on cancer-specific survival was larger also for those with stage III disease.

Discussion

We observed a greater risk of death for people living with IDD and breast (female), colorectal, and lung cancers relative to those without IDD. With few exceptions, worse survival for people with IDD persisted regardless of stage at diagnosis. Our observations are consistent with descriptive studies documenting higher standardized mortality ratios for cancer in people with IDD relative to the general population (Cuypers et al., 2020). Our results are also consistent with multiple scoping reviews detailing inequalities in other cancer care milestones, which support the plausibility of our findings (Boonman et al., 2022; Samtani et al., 2021; Stirling et al., 2021). Our results align with a recent review of cancer care for people living with any disability, which concluded that people with disabilities experienced less treatment and greater cancer mortality than non-disabled people (Iezzoni, 2022).

We hope that our results serve as a call-to-action to research into the causes of worse survival for people with IDD and cancer. Our stage-specific findings of worse

Table 3 Association between intellectual or developmental disability (IDD) status and cancer-specific survival using the cause-specific hazards approach

	N	*N* event (%)	Crude HR (95% CI)	*p*	Adjusted HR[E] (95% CI)	*p*
Breast						
IDD	444	97 (21.8)	2.09 (1.71–2.55)	<0.001	2.28 (1.86–2.78)	<0.001
No IDD	112,331	14,767 (13.1)	Reference		Reference	
Colorectal						
IDD	461	233 (50.5)	2.22 (1.95–2.52)	<0.001	2.57 (2.26–2.92)	<0.001
No IDD	90,980	29,813 (32.8)	Reference		Reference	
Lung						
IDD	355	236 (66.5)	1.28 (1.13–1.46)	<0.001	1.38 (1.21–1.57)	<0.001
No IDD	106,804	70,694 (66.2)	Reference		Reference	

HR hazard ratio, *CI* confidence interval, *IDD* intellectual or developmental disability

[E]Adjusted for age (continuous), rurality, year of diagnosis, geography, sex (*Colorectal* and *Lung*), previous cancer, small-cell cancer (*Lung*), and colon cancer (*Colorectal*)

Table 4 Investigation of effect heterogeneity in the relationship between intellectual or developmental disability (IDD) status and cancer-specific survival by age, sex, and TNM stage

	Breast		Colorectal		Lung	
	Adjusted HR[1] (95% CI)	p^2	Adjusted HR[3] (95% CI)	p^2	Adjusted HR[4] (95% CI)	p^2
Age at diagnosis						
≤49 years	1.89 (1.16–3.10)	0.84	2.92 (2.07–4.13)	0.38	0.89 (0.48–1.66)	0.11
50-59 years	2.15 (1.40–3.31)		2.11 (1.54–2.91)		1.70 (1.26–2.29)	
≥60 years	2.24 (1.74–2.89)		2.33 (2.00–2.72)		1.28 (1.11–1.48)	
Sex						
Male	--	--	2.45 (2.06–2.93)	0.48	1.35 (1.14–1.61)	0.74
Female	--		2.69 (2.23–3.25)		1.42 (1.17–1.72)	
TNM stage						
0/I	0.86 (0.32–2.30)	0.06	2.06 (1.17–3.63)	0.02	1.48 (0.93–2.35)	0.35
II	2.03 (1.36–3.03)		2.10 (1.35–3.26)		1.08 (0.56–2.07)	
III	2.09 (1.39–3.15)		2.60 (1.96–3.44)		1.42 (1.05–1.92)	
IV	1.68 (1.07–2.63)		2.02 (1.63–2.51)		1.23 (1.00–1.52)	
Unknown	3.27 (2.24–4.78)		3.49 (2.74–4.43)		1.67 (1.34–2.08)	

HR hazard ratio, *CI* confidence interval, *IDD* intellectual or developmental disability

1= Adjusted for age, rurality of residence, region of residence, year of cancer diagnosis, previous cancer

2= *p*-value of the interaction term of IDD*age, IDD*sex, IDD*stage calculated using cause-specific Cox proportional hazards regression

3= Adjusted age, sex, rurality of residence, region of residence, year of diagnosis, previous cancer, colon cancer

4= Adjusted age, sex, rurality of residence, region of residence, year of diagnosis, previous cancer, small-cell cancer

cancer-specific survival support the hypothesis that people with IDD may receive less intensive cancer-directed treatment than those without IDD. Even when diagnosed with early-stage cancer where evidence-based curative treatment options exist, we observed that people with IDD experienced a greater risk of death from their cancer. Potential reasons for undertreatment may include concerns from oncologists about non-compliance, challenges in obtaining consent for invasive treatments with significant side effects, and balancing the benefits of the treatment with possible toxicity and side effects (Boonman et al., 2022; Tuffrey-Winje et al., 2013). However, no epidemiological studies have compared receipt of guideline-recommended cancer treatment between people with and without IDD nor does any rigorous scientific evidence indicate adults with IDD experience different preferences for treatment or a higher risk of treatment-related toxicity.

Person-centered strategies within cancer care systems that are adapted to meet the needs of people living with IDD, their families, and their caregivers are required to address these stark inequities. There has been increased awareness of ableism, discrimination against people with disabilities

within the health care system (Janz, 2019). While guidelines emphasize the importance of equity in cancer care (Canadian Partnership Against Cancer, 2019), few if any focus on providing patient-centered care to people living with IDD. Despite primary care clinical guidelines for people living with IDD focusing on facilitating cancer screening (Sullivan et al., 2018), significant disparities in cancer screening persist (Stirling et al., 2021). A different model that engages people with IDD and their caregivers is needed. System-level strategies consisting of education and tailored services co-developed by patients with IDD, their families, and the health care team would strengthen existing structures. Language accessibility training for oncology health care professionals could address challenges with treatment consent and compliance. The integration of specialized nurse or peer navigators for people with IDD could improve coordination between health and community services (NHS Trust, (n.d.)). Additional insurable or publicly funded resources may be required for caregivers of people with IDD through a cancer diagnosis and treatment, including home care.

Our observations should be considered alongside several limitations. Our study documented IDD prevalences of 0.39% among breast (female) cancer patients, 0.51% among colorectal cancer patients, and 0.33% among lung cancer patients, which are lower than estimates in Canada which range from 0.5 to 1.0% of the population. We believe that differences in our cohort likely reflect differences in the risk of cancer for people with IDD (e.g., if rates of smoking are lower among people with IDD, their prevalence among a population with a smoking-related cancer would be smaller), and differences in the age distributions used in reporting prevalence (e.g., if people with IDD do not live long enough to develop cancer, the prevalence would be higher among younger age populations) and in the data sources used in our study. Our algorithm to identify people with IDD is not validated and does not include non-health information sources. Misclassification likely occurs among younger adults with IDD and those with fewer health care system encounters (Lin et al., 2013) and should not change our conclusions. Our definition of IDD groups together multiple diagnoses and severities of disability. The risk of dying from cancer is likely higher for those with the most severe features, as this group may experience restrictions that most interfere with screening and treatment decision-making. Loss to follow-up due to moving out of the province is not a variable captured in the ICES data. However, there are no data suggesting people with IDD are more or less likely to move out of province following a cancer diagnosis and therefore we do not anticipate that out of province migration would change the conclusions of the study. Finally, the proportional hazards assumption was violated. While on average the risk of dying was greater for people with IDD relative to those without during the study period, the size of differences differed statistically over time. We expect this occurred due to the large sample size, the extended follow-up period, the significant interaction with stage at diagnosis, and a greater risk of dying immediately following the cancer diagnosis due to a lack of curative treatment. However, this does not change our conclusions. For example, in the lung cancer cohort, the HR in the first 2 years and after 2 years were 1.39 and 1.30, compared with 1.37 for the overall average.

Future research should include studies of cancer survival for people with specific diagnoses or disabilities and conducted in other jurisdictions to confirm our results. Studies of other cancers, in particular those with higher rates of mortality among those with IDD such as digestive organs besides colon, bladder, and cervix cancer (Cuypers et al., 2020), would be beneficial. Research anchored in the principles of intersectionality within this population is also needed (Carbado et al., 2013).

Conclusion

This study provides foundational population-based evidence of cancer survival inequities experienced by adults living with intellectual or developmental disabilities. High-quality knowledge to inform patient-centered, evidence-based care gaps for people with IDD and cancer is urgently needed. This includes initiating ongoing surveillance of cancer survival disparities for people with IDD to ensure efforts result in survival gains and benefits, as well as studies identifying, measuring, and intervening on cancer outcomes prioritized by people with IDD. The balance of existing evidence is adequate to necessitate action and advocacy now. People with IDD diagnosed with cancer are equally deserving of opportunities to survive their cancer diagnosis as those without IDD.

Contributions to knowledge

What does this study add to existing knowledge?

- Although cancer is a leading cause of death among people with intellectual or developmental disabilities (IDD), few studies have examined cancer survival. The few that exist employ lower-quality study designs, such as using insufficient data sources or not including a general cancer population comparator group.
- Our study compared breast (female), colorectal, and lung cancer survival for adults with IDD to those without IDD using administrative health data from Ontario. We documented significantly worse cancer survival for people with IDD across each cancer type. With few exceptions, worse survival for people with IDD persisted regardless of stage at diagnosis.

What are the key implications for public health interventions, practice, or policy?

- There are few adequate, ethical explanations for worse cancer survival for people living with IDD and many opportunities to increase the provision of patient-centered oncology care exist.
- Developing high-quality scientific evidence to inform oncology practice and to develop patient-centered strategies to improve cancer survival for people with IDD above and beyond increasing cancer prevention and screening efforts is paramount.
- High-quality research describing and comparing receipt of guideline-recommended treatment is needed to inform people with IDD, their families, and their health care teams to ensure curative options are offered and considered.

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Author contributions Hansford: literature search, figures, data interpretation, writing–original draft, writing–reviewing and editing; Ouellette-Kuntz, Cobigo: conceptualization, funding acquisition, methodology, data interpretation, writing–reviewing and editing; Griffiths: methodology, formal analysis, writing–reviewing and editing; Hallet, Decker, Dawe, Kristjanson, Shooshtari, Stirling, Brownell, Turner: funding acquisition, methodology, data interpretation, writing–reviewing and editing; Kelly: data interpretation, writing–reviewing and editing; Mahar: conceptualization, funding acquisition, methodology, data interpretation, writing–original draft, writing–reviewing and editing, supervision.

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Availability of data and material, and code availability The dataset used in this study is held securely in coded format at ICES. ICES is a prescribed entity under section 45 of Ontario's Personal Health Information Protection Act. Section 45 authorizes ICES to collect personal health information, without consent, for the purpose of analysis of compiling statistical information with respect to the management of, evaluation or monitoring of, the allocation of resources to, or planning for all or part of the health system. Legal restrictions and data sharing agreements prohibit ICES from making the dataset publicly available. Access may be granted to those who meet the conditions for confidential access, available at https://www.ices.on.ca/DAS. AM and HOK hold appointments as ICES Scientists, which enabled access to ICES data. Data access is available to external public sector researchers either through collaboration with an ICES scientist or directly, following project approval, via a secure online desktop infrastructure (see above link for details).

Declarations

Conflict of interest RH, HOK, RG, KD, MK, VC, SS, MS, CK, MB, DT, and ALM declare no competing interests. JH has received payments for speaking honoraria from Ipsen Biopharmaceuticals, Advanced Accelerator Applications, Medtronic, and Brystol-Myers-Squibb. DED has received research grants from AstraZeneca, CIHR, CancerCare Manitoba Foundation, and the Manitoba Medical Services Foundation, as well as honoraria for education materials from Boehringer-Ingelheim and Bristol-Myers Squibb and participates as an advisory board member for AstraZeneca, Merck Canada, Jazz Pharmaceuticals, Pfizer, and Novartis. DED is a member of the Lung Cancer Canada Medical Advisory Committee, is a provincial representative on the System Performance Group for the Canadian Partnership Against Cancer, and the Medical Lead for the Frail and Older Adult initiative at CancerCare Manitoba.

Ethics approval The study received ethical clearance from the University of Manitoba Health Research Ethics Board (#H2019:253) and Queen's University Faculty of Health Sciences and Affiliated Hospitals Research Ethics Board (#EPID-691-19).

Consent to participate Not applicable.

Consent for publication Not applicable.

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References

Austin, P. C. (2009). Using the standardized difference to compare the prevalence of a binary variable between two groups in observational research. *Communication in Statistics- Simulation and Computation, 38*, 1228–1234.

Austin, P. C., Lee, D. S., & Fine, J. P. (2016). Introduction to the analysis of survival data in the presence of competing risks. *Circulation, 133*, 601–609.

Boonman, A. J., Cuypers, M., Leusink, G. L., Nalldenberg, J., & Bloemendal, H. J. (2022). Cancer treatment and decision making in individuals with intellectual disabilities: A scoping literature review. *Lancet Oncology, 23*, e174-183.

Brenner, D. R., Poirier, A. E., Woods, R. R., Ellison, L. F., Billette, J. M., Demers, A. A., Zhang, S. X., Yao, C., Finley, C., Fitzgerald, N., Saint-Jacques, N., Shack, L., Turner, D., & Holmes, E. (2022). Projected estimates of cancer in Canada in 2022. *CMAJ, 194*, E601-607. https://doi.org/10.1503/cmaj.212097

Canadian Partnership Against Cancer. (2019). *Canadian strategy for cancer control: Doing together what cannot be done alone*. https://s22457.pcdn.co/wp-content/uploads/2019/06/Canadian-Strategy-Cancer-Control-2019-2029-EN.pdf. Accessed 15 Jun 2022.

Carbado, D. W., Crenshaw, K. W., Mays, V. M., & Tomlinson, B. (2013). Intersectionality: Mapping the movements of a theory. *Du Bois Review, 10*(2), 303–312. https://doi.org/10.1017/S1742058X13000349

Clarke, E. A., Marrett, L. D., & Kreiger, N. (1991). Cancer registration in Ontario: A computer approach. In *Cancer registration principles and methods.* (pp. p. 246-257). IARC.

Cuypers, M., Schalk, B. W. M., Boonman, A. J. N., Nalldenberg, J., & Leusink, G. L. (2020). Cancer-related mortality among people with intellectual disabilities: A nationwide population-based cohort study. *American Cancer Society, 128*, 1267–1274. https://doi.org/10.1002/cncr.34030

Glover, G., Williams, R., Heslop, P., Oyinlola, J., & Grey, J. (2017). Mortality in people with intellectual disabilities in England. *Journal of Intellectual Disability Research, 61*, 62–74. https://doi.org/10.1111/jir.12314

Goel, R., & Beder, M. (2012). Welcome to Canada … but don't get sick. *CMAJ, 184*(1), E103. https://doi.org/10.1503/cmaj.111095

Hess, K. R. (1993). Graphical methods for assessing violations of the proportional hazards assumption in Cox regression. *Statistics in Medicine, 14*, 1707–1723.

Iezzoni, L. I. (2022). Cancer detection, diagnosis, and treatment for adults with disabilities. *Lancet Oncology*, *23*(4), e164–e173.

Jackson, J. W. (2021). Meaningful causal decompositions in health equity research: Definition, identification, and estimation through a weighting framework. *Epidemiology, 32*, 282–290. https://doi.org/10.1097/EDE.0000000000001319

Janz, H. L. (2019). Ableism: The undiagnosed malady afflicting medicine. *Canadian Medical Association Journal, 191*, E478-479. https://doi.org/10.1503/cmaj.180903

Johns Hopkins University. (2014). The Johns Hopkins ACG® System Version 11.0 Technical Reference Guide. Johns Hopkins Bloomberg School of Public Health. Baltimore, MD.

Krahn, G. L., Hammond, L., & Turner, A. (2006). A cascade of disparities: Health and health care access for people with intellectual disabilities. *Mental Retardation and Developmental Disabilities, 12*, 70–82.

Kralj, B. (2009). *Measuring rurality- RIO 2008_Basic: Methodology and results*. Ontario Medical Association Economics Department.

Lin, E., Balogh, R., Cobigo, V., Ouellette-Kuntz, H., Wilton, A. S., & Lunsky, Y. (2013). Using administrative health data to identify individuals with intellectual and developmental disabilities: A comparison of algorithms. *Journal of Intellectual Disability Research, 57*, 462–477. https://doi.org/10.1111/jir.12002

Maulik, P. K., Mascarenhas, M. N., Mathers, C. D., Dua, T., & Saxena, S. (2011). Prevalence of intellectual disability: A meta-analysis of population-based studies. *Research in Developmental Disabilities*. https://doi.org/10.1016/j.ridd.2010.12.018

NHS Trust. (n.d.). *Help for people with learning disabilities and autism*. https://www.bhrhospitals.nhs.uk/help-for-people-with-learning-disabilities/. Accessed 15 Jun 2022.

Ouellette-Kuntz, H. (2005). Understanding health disparities and inequities faced by individuals with intellectual disabilities. *Journal of Applied Research in Intellectual Disabilities, 18*, 113–121.

Ouellette-Kuntz, H., & Lynn, M. (2014). *Applied health research question report: Aging profiles of adults with and without developmental disabilities in Ontario*. https://www.porticonetwork.ca/documents/38160/99698/Aging_AHRQ+report/ac2fb8e7-5a80-457c-913a-7279fd5733b7. Accessed 15 Jun 2022.

Patja, K., Iivanainen, M., Vesala, H., Oksanen, H., & Ruoppila, I. (2000). Life expectancy of people with intellectual disability: A 35-year follow-up study. *Journal of Intellectual Disability Research, 44*(Pt 5), 591–599.

Robles, S. C., Marrett, L. D., Clarke, E. A., & Risch, H. A. (1988). An application of capture-recapture methods to the estimation of completeness of cancer registration. *Journal of Clinical Epidemiology, 41*(5), 495–501.

Samtani, G., Bassford, T. L., Williamson, H. J., & Armin, J. S. (2021). Are researchers addressing cancer treatment and survivorship among people with intellectual and developmental disabilities in the U.S.? A scoping review. *Intellectual and Developmental Disabilities, 59*, 141–154. https://doi.org/10.1352/1934-9556-59.2.141

Stankiewicz, E., Ouellette-Kuntz, H., McIsaac, M., Shooshtari, S., & Balogh, R. (2018). Patterns of mortality among adults with intellectual and developmental disabilities in Ontario. *Canadian Journal of Public Health, 109*, 866–872. https://doi.org/10.17269/s41997-018-0124-8

Statistics Canada. (2022). *Population estimates, quarterly*. https://www150.statcan.gc.ca/t1/tbl1/en/tv.action?pid=1710000901. Accessed 15 Jun 2022.

Stirling, M., Anderson, A., Ouellette-Kuntz, H., Hallet, J., Kelly, C., Dawe, D. E., Kristjanson, M., Decker, K., & Mahar, A. L. (2021). A scoping review documenting cancer outcomes and inequities for adults living with intellectual and/or developmental disabilities. *European Journal of Oncology Nursing*, *54*. https://doi.org/10.1016/j.ejon.2021.102011

Sullivan, W. F., Diepstra, H., Heng, J., Ally, S., Bradley, E., Casson, I., Hennen B., Kelly, M., Korossy, M., McNeil, K., Abells, D., Amaria, K. , Boyd, K., Gemmill, M., Grier, E., Kennie-Kaulbach, N., Ketchell, M., Ladouceur, J., Lepp, A., Lunsky, Y., McMillan, S., Niel, U., . . . Witherbee, S. (2018). Primary care of adults with intellectual and developmental disabilities: 2018 Canadian consensus guidelines. *Canadian Family Physician*, *64*, 54–79.

Tuffrey-Winje, I., Nikoletta, G., Butler, G., Cresswell, A., Manners, P., & Bernal, J. (2013). Developing guidelines for disclosure or non-disclosure of bad news around life-limiting illness and death to people with intellectual disabilities. *Journal of Applied Research in Intellectual Disabilities, 26*, 231–242. https://doi.org/10.1111/jar.12026

von Elm, E., Altman, D. G., Egger, M., Pocock, S. J., Gøtzsche, P. C., & Vandenbroucke, J. P. (2007). The Strengthening the Reporting of Observational Studies in Epidemiology (STROBE) statement: Guidelines for reporting observational studies. *PLoS Med, 4*(10), e296. https://doi.org/10.1371/journal.pmed.0040296