An invisible population: late-stage cancer diagnosis for people with intellectual and developmental disability

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Abstract

In this issue of *Cancer*, Mahar and colleagues report stark findings that people with IDD experienced a 60% higher risk of stage IV female breast cancer, and a 42% higher risk of stage IV colorectal cancer, compared to those without IDD. With no comparable studies in the literature, the findings have international research, practical and clinical relevance highlighting health inequities in access to curative-intent cancer treatments for a population not well-identified, and often voiceless in health systems that are difficult to navigate.

Intellectual or developmental disabilities (IDD) affects over 1.4% of the global population and includes delays and limitations in functioning that manifest during the developmental period as a result of disorders or injuries affecting neurodevelopment.(1) People with IDD are at a significantly greater risk than their peers from common social determinants of health(2) associated with increased risk of developing cancer(3) and poorer health outcomes.(4) Individual factors including health and lifestyle behaviours, as well as structural and institutional factors (for example, the way health systems are organised and delivered) put people with IDD at greater risk of developing cancer, receiving a late diagnosis and experiencing poorer outcomes. (2, 5)

As the authors also note, people with IDD experience greater levels of poverty,[6] unemployment,[7] social isolation,[8] and many live in precarious living environments. [7] People with IDD have high rates of obesity and sedentary behaviour,[9] have an atypical pattern of healthcare utilisation[10] and have high rates of physical and mental health issues.[11-13] These factors are associated with increased cancer risk and also limit the ability of people with IDD to receive timely diagnosis and optimal cancer care.

Mahar et al. highlight that even though there were organised population-based cancer screening programs available, people with IDD still presented at a later stage for breast (female) and colorectal cancer compared to those without IDD. This is not unique to Canada. Recent evidence from England not only concurs,[14] but highlights that among cancer deaths in people with IDD (N=1,096), of those who died with colorectal cancer, 43% were below the age threshold for colorectal screening. Similar opportunities for people with IDD to avail of the national screening program, did not reduce inequities. With such a high percentage dying before reaching the screening age threshold, this highlights that screening criteria were not addressing the needs of people with IDD. The Mahar et al study finding regarding higher levels of late-stage diagnosis for breast (female) and colorectal cancers again point out that age criteria for screening designed for the general population may not be adequate or appropriate for people with IDD.

Mahar and colleagues rightly suggest that extended surveillance and early intervention for cancer among individuals with IDD is warranted. This is a worthy recommendation deserving of meaningful attention. However, it is well documented that as people with IDD leave school-based programmes and become adults, whether by choice or exclusion many people with IDD rapidly become unknown to health and social care services with a marked discrepancy between the 'administrative' and 'true' prevalence of IDD. This is often referred to as the 'visible minority', i.e., those known to health and social care services and the 'hidden majority', i.e. those with IDD but not known to health and social care services.[2] In the IDD literature, this is referred to as the 'transition cliff'[2] where children with IDD reaching adulthood become "lost" and whose lives are less likely to be identified and followed making the construction and monitoring of life and health histories more difficult and unlikely.

Longitudinal analysis as compared to cross sectional analysis confirms this trend. For example, a longitudinal study on ageing and intellectual disability (IDS-TILDA) in Ireland reports high screening uptake for colorectal and breast cancers in comparison to the general population;[15] however, larger administrative studies of people with IDD in Canada [16] and Denmark[17] report lower screening uptake for these cancers. The differences likely speak to this issue that where purposeful follow-up is absent, there are a subgroup of adults with IDD who are not known to any services, the 'hidden majority' of people with IDD, and they may be vulnerable to not being screened early enough or frequently enough leading to a greater likelihood of late-stage cancer diagnosis.

As noted by Maher and colleagues late-stage cancer diagnosis limits opportunities for curative treatment which then impacts survival rates. Such concerns are further compounded by findings in a recent review that upon diagnosis people with IDD receive less intensive cancer treatment than is generally administered to other populations. [18] This is an additional inequity. The principles for treating cancer should be the same for people with IDD as the general population and they should receive the most effective care and treatment. In practice, such delivery of care can be complicated by the severity of the person's IDD, their communication ability, caregiver influence, complex ethical and legal considerations, diagnostic and treatment challenges where sedation and anaesthesia may be required, behavioural presentations and absence of the capacity to consent. These factors pose a real ethical and moral dilemma, which resolves around the challenges of disentangling the advantages and disadvantage of a treatment approach while striving to maintain a balance between outcomes and quality of life for people with IDD.

Mahar et al. have a contrasting finding that people with and without IDD were as likely to be diagnosed with stage IV lung Cancer. The authors do highlight the higher rates of missing stage data among those with IDD and lung cancer compared to those without, but it is important to underline Mahar et al.'s point that that rates of tobacco smoking may also be relevant. In the field of IDD there are conflicting reports on whether tobacco smoking is higher or lower in this population.[19] Perhaps lower in the 'visible minority', it may be higher in the 'hidden majority' of people with IDD who often have a lower socioeconomic status but it remains to be fully researched as does the prevalence of exposure to carcinogens associated with lung cancer development. However, Mahar and colleagues most important and actionable findings are in terms of breast and colorectal cancers.

Action requires first, the 'transition cliff' which can also involve a disconnect from health services must be mitigated if screening and health promotion programs are to be most effectively offered to people with IDD. That people with IDD present with greater risks of developing these cancers and are more likely to receive later stage cancer diagnosis will then be more likely to be addressed. Primary care physicians and specialist community services play a leading role in pre-emptive health surveillance (often referred to as an Annual Health check in the UK) and a second priority is that they must increase their engagement in regular checks and evaluation for symptoms or signs of illness in people with IDD. Greater attention and knowledge about people with IDD and their risks for these cancers will then mean that resources may be planned and delivered at a scale and intensity proportionate to the substantial and wide-ranging degree of need among people with IDD.[4] Third, to advance such surveillance and action, professional health care training programs across disciplines must better address understanding of IDD and health responses/non-responses that negatively impact the standard of care, and diagnosis and treatment delivery. As Mahar et al. cite healthcare providers are too often influenced by 'diagnostic overshadowing' and 'ableism'. Fourth, reasonable adjustments, the positive changes made to accommodate the needs of people with IDD need to be instituted across all health services including increasing length of appointments, making information accessible, involving people with IDD in decision making at an earlier stage, and developing advanced care directives and health or hospital passports documenting a person's ability, needs, general health and desires for treatment.

Perhaps most critical is a fourth recommendation that some cancer screening programs may need to be available at an earlier age for this population; however, rigorous empirical work needs to be undertaken to justify this first.

Finally, research in the area of cancer and IDD is lacking. While large scale administrative studies are important to identify and detect associations at a population level, prospective cohort studies will better assess which specific IDD determinants (lifestyle and genetic factors, behaviours, environmental exposures, and healthcare utilisation) are associated with increased cancer risk and poorer outcomes. Such studies are costly and difficult to coordinate, and an international approach is needed to unite resources. In Europe, a recent COST Action 'Cancer- Understanding Prevention in Intellectual Disabilities' (CUPID)[20] project is developing a research agenda and knowledge base to improve cancer care

inequalities for people with IDD in the European Union and beyond. Mahar's et al. findings will inform the direction of this COST Action and improve the likelihood that future research will signpost how to reduce inequities in cancer care that people with IDD experience.

Most countries are signatory to the UNCRPD protocol (2006) which sets out under Article 25a that people with disabilities should be provided with *"...the same range, quality and standard of free or affordable health care…*". From this viewpoint, the inequity reported in Mahar et al's study needs to be addressed as a matter of priority.

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