

Rare but Not Uncommon: Rare Cancers in North America

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In North America, the most common sites in which cancer develops include the breast, prostate, lung, bowel, bladder, and uterus.¹ These cancers comprise approximately 55% of all cancers diagnosed in North America.¹ Yet, there are 100s of different types of cancer. Other cancers, such as 'rare' cancers, collectively contribute a significant share of the cancer burden. Of all cancers diagnosed in the United States from 2009 to 2013, one fifth were considered rare.² A recent study reported that from 2006 to 2016, there were 154,900 rare cancer diagnoses made in Canada, meaning that 17% of all cancers diagnosed were rare cancer diagnoses.³

What is a rare cancer?

While there is no universally accepted definition, a measure of fewer than six cases per 100,000 individuals per year was put forth by the Surveillance of Rare Cancer in Europe⁴ and has been recently used in practice.^{2,3,5} For comparison, the most diagnosed cancer in Canada, lung cancer, had an estimated 68 cases per 100,000 people in 2020.⁵ The definition of fewer than six cases per 100,000 individuals per year means that most cancers among children and adolescents are rare cancers. For example, in the United States, 71% of cancers diagnosed in children and adolescents are considered rare cancers.²

Cancers are classified according to body site (i.e., breast, lung, etc.) where the cancer first develops and the type of tissue it first develops in (referred to as histological type). Cancers can be classified according to hundreds of different histologies. Although leukemia is not rare, a specific histology of leukemia called myeloid leukemia is considered a rare cancer because there are fewer than 6 cases diagnosed per 100,000 individuals per year. Using data collected from 2009 to 2013, it is estimated that there are 181 types of rare cancers in the United States, of which 119 are very rare with 0.5 or fewer cases per 100,000.²

What challenges do rare cancers pose?

Since few patients have any given rare cancer, developing and testing possible treatments for those cancers can be more challenging than it is for other more common cancers. For example, it can be difficult to recruit participants into a clinical trial and even to obtain tumour tissue to study.⁷ Since rare cancers can also be more difficult to diagnose, misdiagnoses and delays are more common.^{2,7,8} Specifically, doctors may have less knowledge of rare cancers⁷ and may also find it difficult to identify an expert in a specific rare cancer to refer their patient to. The 5-year relative survival for rare cancers was worse than common cancers for males (55% versus 75%) and females (60% versus 74%).² The survival difference is largely attributed to the relatively later stage that rare cancers get diagnosed.² Despite the cascade of challenges that rare cancers pose to researchers, doctors and most importantly, patients, advocacy groups have made great strides in supporting research on how to better diagnose and treat rare cancers.

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