Background

There is no universally adopted definition for rare cancer:

- The US Orphan Drug Act of 1983 defined rare diseases as those affecting less than 200,000 people in the United States.
- In 2010, Greenlee et al described the US burden of rare cancers according to the National Cancer Institute’s definition as those cancers with fewer than 15 cases per 100,000 people per year.\(^1\)
- More recently, a consortium from the European Union, Surveillance of Rare Cancer in Europe (RARECARE), described the burden of rare cancers in Europe using a revised definition of rare cancers as those with fewer than 6 cases per 100,000 people per year.\(^2\)

Rare cancers present unique challenges for clinicians and their patients:

- For most rare cancers, research to identify causes or to develop strategies for prevention or early detection is limited or nonexistent.
- In addition, rare cancers can be challenging to diagnose, often resulting in numerous physician visits, misdiagnoses, and substantial delays in diagnosis.
- Treatment options for rare cancers are often more limited and less effective than for more common cancers.

References:


Methods

- Rare cancers were defined according to the RARECARE cancer list as those with an incidence fewer than 6 cases per 100,000 individuals per year.
- Some cancers with non-specific histology and/or morphology codes could not be classified as rare or common.
- Incidence rates and case distributions by sex, race/ethnicity, age, and stage at diagnosis were obtained using North American Association of Central Cancer Registries (NAACCR) data from 47 states and the District of Columbia for patients diagnosed during 2009-2013.
- Data for survival analyses were obtained from the Surveillance, Epidemiology, and End Results (SEER) program of the National Cancer Institute SEER 18 registries. One-year and 5-year relative survival were based on patients who were diagnosed during 2008-2012 and 2006-2012, respectively, and were followed through 2013.

Results

Figure 1. Distribution of Rare and Common Cancers, 2009-2013, United States

Figure 2. Late-stage Distribution for Rare and Common Cancers, 2009-2013, United States

Figure 3. Five-year Relative Survival for Rare and Common Cancers by Age at Diagnosis (years), 2006-2012, United States

Figure 4. Five-year Relative Survival for Rare and Common Cancers by Age at Diagnosis (years), 2006-2012, United States

Conclusions

- Approximately 1 in 5 cancer patients in the United States are diagnosed with a rare cancer. A larger proportion of cancers in children are rare compared to older adults.
- Among solid tumors, 59% of rare cancers are diagnosed at regional or distant stages compared to 45% of common cancers.
- In part because of this stage distribution, 5-year relative survival is lower for patients with a rare cancer than those diagnosed with a common cancer among both males (55% vs 75%) and females (60% vs 74%).
- However, 5-year survival is higher for children and adolescents diagnosed with a rare cancer (82%) than adults (46% for ages 65-79). The advancement of treatment of many childhood cancer is due in large part to concentration of treatment in specialized centers and the joint research collaboration among these centers. Similar collaborations are needed to advance treatment for rare cancer in adults.
- Continued efforts are needed to develop interventions for prevention, early detection, and treatment to reduce the burden of rare cancers. Discoveries for rare cancers can further knowledge for all cancers.