

## **Transcript - JAMA Ophthalmology interview (4-10-14) with Jeannette Wong on Retinoblastoma Incidence Patterns in the United States**

Deanna Bellandi: [Music] Hello and welcome to this author interview from the JAMA Network. This is Deanna Bellandi with JAMA Ophthalmology. Several studies have found no differences in the incidence of retinoblastoma, a rare childhood cancer, except for age at diagnosis, but other studies have reported differences in incidence by sex and race and ethnicity. Jeannette Wong of the National Cancer Institute at the National Institutes of Health and colleagues examined U.S. retinoblastoma incidence patterns by sex, age at diagnosis, race, ethnicity and year of diagnosis. Welcome, Ms. Wong.

Jeannette Wong: Thank you for having me.

Deanna Bellandi: So can you tell me a little bit more about what you studied?

Jeannette Wong: We set out to characterize retinoblastoma incidence patterns in the United States. Our study included retinoblastoma cases diagnosed before the age of 5 between the years 1975 and 2009 from 18 United States cancer registries. Our study allowed the largest number of available cases from this database to be included and allowed us to explore overall incidence patterns, and for the first time characterize patterns by sex, age at diagnosis, laterality and by race and ethnicity.

Deanna Bellandi: So what did you find?

Jeannette Wong: Overall, we found retinoblastoma incidence rates in the United States did not change dramatically. The retinoblastoma incidence rate in the recent decade was almost 13 cases per million per year. We were also able to explore population differences by a variety of demographic factors for the first time, but since these analyses were based on small numbers, we're keen to follow up on this study as more case data becomes available in the future.

Deanna Bellandi: So what do the results mean?

Jeannette Wong: So our results provide an important baseline assessment for understanding retinoblastoma incidence in the United States. Retinoblastoma is a rare childhood cancer and it accounts for 11 percent of cancers occurring in the first year of life, so it is important for us to establish an understanding of retinoblastoma now to be able to continue following this as more data becomes available.

Deanna Bellandi: Can you discuss some of the trends in your findings?

Jeannette Wong: So we saw some differences by gender and we noticed a recently emerging male predominance retinoblastoma and also some population differences by race and ethnicity. We were specifically able to report patterns and trends for Hispanic ethnicity and Asian Pacific Islander populations for the first time. Our results are based on small numbers and so the estimates we reported in our study could be due to chance or perhaps unstable estimates and we hope to provide more stable estimates and validation to what we're reporting for the first time.

Deanna Bellandi: So what are the implications for patients and clinicians because of what you found?

Jeannette Wong: I'm not a clinician but I think some things that are important to remember is that we are dealing with a rare childhood cancer and while overall the retinoblastoma incidence rates have not changed dramatically, survival rates for retinoblastoma have greatly improved. With the five year survival rate of almost 97 percent it is important to consider the long term health impacts to retinoblastoma survivors and how studying these survivors can help us understand retinoblastoma even more. We know from several studies that retinoblastoma survivors, especially those with the germline mutation in the retinoblastoma gene, are at an increased risk for developing second cancers. There are also survivors who do not have a germline mutation but rather, de novo mutation in the retinoblastoma gene, and we don't know if there are environmental factors for retinoblastoma and there currently are no known environmental factors, but these retinoblastoma cases from de novo mutations do persist. And so using those

survivors with de novo and germline mutations could provide new insight into understanding the etiologic factors related to retinoblastoma development.

Deanna Bellandi: What impact if any did prenatal screening for retinoblastoma have on the incidence numbers?

Jeannette Wong: So our exploratory analysis looking at incidence trends over time revealed a correlation between calendar period of diagnosis and when genetic screening became available for retinoblastoma. However there is no comprehensive data on the use of genetic testing or screening for retinoblastoma but hopefully with more comprehensive medical care in the future we can explore this question a little bit more. Our study does not impact use of genetic screening for retinoblastoma since the study itself focuses on the trends in the United States overall for retinoblastoma incidence.

Deanna Bellandi: So what are the next steps in terms of research?

Jeannette Wong: So again this goes back to the idea of looking at those retinoblastoma survivors who have germline mutations versus de novo mutations and really utilizing the survivor populations to understand the etiologic factors behind retinoblastoma development a little bit more. We certainly know a lot more about some of the—for example—second cancers that happen in retinoblastoma survivor populations, but now that we've provided this comprehensive perspective on retinoblastoma trends in the United States, it would be useful to see if we can use these retinoblastoma survivors to explore underlying risk factors for retinoblastoma as well.

Deanna Bellandi: [Music] Thank you Ms. Wong for talking to us about your study. This is Deanna Bellandi with JAMA Ophthalmology. For more podcasts visit us online at [JAMA.com](http://JAMA.com).