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## Evaluation of retinoblastoma incidence in children in Los Angeles and Ventura counties

### Introduction

On May 21<sup>st</sup> 2007, Dr. Margaret McCusker, Chief of the Cancer Surveillance Section, received a request from the office of Senator Sheila Kuehl to review the incidence of retinoblastoma in Los Angeles and Ventura Counties, with a focus on the area around the Santa Susana Field Laboratory (SSFL). There was a community concern that the risk of retinoblastoma (RB) was increased in children as a result of potential cancer-causing contaminants in the vicinity of SSFL. Senator Kuehl asked the Cancer Surveillance Section to update a 2005 analysis conducted by the University of Southern California (USC) Cancer Surveillance Program that included cases diagnosed through 2002 and showed no excess incidence of retinoblastoma in this area.

### Methods

Following the standard California Cancer Registry (CCR) protocol for assessing cancer concerns, CCR research staff assessed whether the number of cases diagnosed in the area around the SSFL was greater than would be expected based on overall incidence in California. An analysis plan was developed in consultation with Dr. Hal Morgenstern, an epidemiologist at the University of Michigan with an interest in this topic. The area of interest was defined as a ten mile radius around the SSFL, and the census tracts located in this area were identified (see attached map). The area encompassed the communities of Simi Valley, Oak Park, and part of West Lake Village in Ventura County; and Chatsworth, Canoga Park, Woodland Hills,

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Calabasas and Agoura in Los Angeles County. The time period of interest was 1988 (the beginning of the statewide cancer registry) through 2005 (the most recent year of complete data). Because 95% of all RB patients are diagnosed before age 5, cases for this analysis were defined as children under the age of 5 with RB. All retinoblastoma cases (ICDO2 9510-9512 and ICDO3 9510-9514) diagnosed between 1988 and 2005 in children under age 5 were included in the analysis. The reference population was all of California, and the expected number of cases was calculated by multiplying the race- and sex-specific rates of retinoblastoma in children under age 5 in California by the race- and sex- population under age 5 in the area of interest. Because the time period of interest encompassed both the 1990 and the 2000 census, the 1990 census population was used to calculate the expected values for 1988-1995; and the 2000 census was used to calculate the expected values for 1996-2005. The population of children under age 5 in the combined area was 29,331 in 1990, and 31,601 in 2000.

## Results

Based on the incidence of retinoblastoma among children under age 5 in California, and the population under age 5 in the SSFL area, the number of cases that would be expected to occur in this area between 1988 and 2005 was calculated to be 7.5. The total number of retinoblastoma cases observed in this area between 1988 and 2005 was 11, with a 99% confidence interval of 4.3-22.8 cases. Since the confidence interval around the observed included the expected number, the incidence of retinoblastoma in the area of interest was not statistically significantly elevated.

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The age distribution of the 11 cases was: 5 under age 1, 1 age 1, 3 age 2, 1 age 3, 1 age 4. Five of the cases had bilateral disease and 6 had unilateral disease. Nearly half (5 cases) were Hispanic, the remainder were non-Hispanic white (n=3), non-Hispanic black (n=1), non-Hispanic Asian/Pacific Islander (n=1), and unknown race/ethnicity (n=1). All 11 cases were diagnosed in 1998 or later: 1 in 1998, 1 in 1999, 1 in 2000, 1 in 2001, 2 in 2002, 1 in 2003, 3 in 2004, and 1 in 2005. All cases resided in the three neighborhoods with the largest populations (Canoga Park, Woodland Hills, Simi Valley) but there were too few cases to analyze incidence in individual neighborhoods.

## Discussion

Approximately half of retinoblastomas are believed to be genetic in origin. The hereditary form of retinoblastoma is characterized by earlier age at onset and increased probability of bilateral disease. Each child of a parent with familial bilateral retinoblastoma has a 50% risk of inheriting the retinoblastoma gene. Children who inherit the retinoblastoma gene have a 90% risk of developing retinoblastoma. Little is known about non-genetic risk factors for this disease, although one study suggested that parental occupation in the military or in metal manufacturing was associated with retinoblastoma.

According to the literature, approximately 25% of retinoblastoma cases are bilateral, thus the nearly half of bilateral cases in this population was higher than expected. Since bilateral disease is heritable, it is possible that the children in this analysis with bilateral disease have a family history of retinoblastoma. A positive family history was demonstrated in the registry abstract of

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one of the cases. Nearly half of the cases diagnosed in this population were diagnosed among children under one year of age, also an indication of the hereditary form of the disease. Half of the cases diagnosed in the study area were Hispanic, while only 19% of the population of the area of interest under age 5 is Hispanic. Incidence of retinoblastoma is higher among Hispanics and blacks in California.

## Conclusion

In conclusion, incidence of retinoblastoma among children under age 5 residing in the area around the SSFL between 1988 and 2005 was slightly, although not statistically significantly, higher than expected based on incidence statewide. The relatively young age of the cases, and the high proportion of cases with bilateral disease, is suggestive of a genetic origin. This analysis is consistent with the 2005 report that showed no significant increased risk of retinoblastoma between 1972 and 2002.