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## Incidence Patterns of Adrenocortical Carcinomas and Malignant Pheochromocytomas and Paragangliomas in California

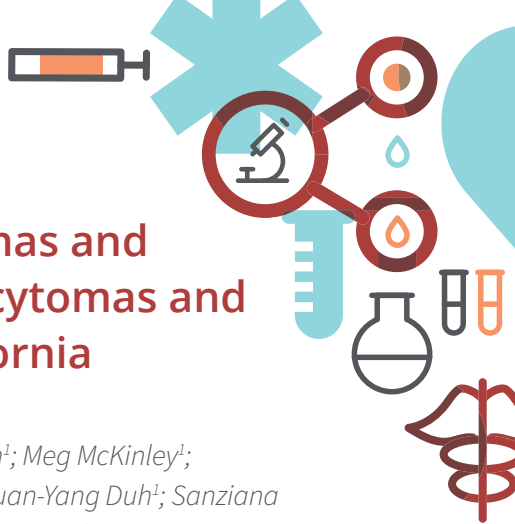
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**BACKGROUND:** Pheochromocytomas (PHEO), paragangliomas (PGL), and adrenocortical carcinomas (ACC) are rare endocrine malignancies with limited data regarding risk factors. We sought to characterize the burden of malignant PHEO, PGL, and ACC in California.

**METHODS:** Using the population-based California Cancer Registry, we identified all new diagnoses of malignant PHEO, PGL, and ACC in California from 1992-2016 (ICD-O-3 codes 8700/3, 8680/3, and 8393/3). We estimated age-adjusted incidence rates (AIR) standardized to the 2000 United States census. We compared AIRs by sex, race/ethnicity, and county of residence categorized as urban versus suburban/rural using SEER\*Stat and generated IR ratios (IRR).

**RESULTS:** Between 1992-2016, there were 261 incident cases of malignant PHEO, 271 of malignant PGL, and 866 of ACC in California. Overall AIR per 100,000 person-years were 0.03 for PHEO, 0.025 for PGL, and 0.10 for ACC. Incidence differed by both sex and race/ethnicity (Table 1). Men had a higher incidence of PGL than women ( $p < 0.05$ ) and tended towards higher incidence of PHEO ( $p = 0.09$ ), while men had a lower incidence of ACC than women ( $p < 0.05$ ). Compared with non-Hispanic White Californians, non-Hispanic Black Californians had higher PHEO incidence, whereas PGL incidence was lower for



Hispanic and Asian/Pacific Islander Californians ( $p < 0.05$ ). Compared with non-Hispanic White Californians, ACC incidence was lower in non-Hispanic Black, Hispanic, and Asian/Pacific Islander Californians (all  $p < 0.05$ ). There were no differences in incidence of either PHEO, PGL, or ACC for urban versus suburban/rural counties.

**CONCLUSION:** Malignant PHEO, PGL, and ACC remain rare cancers in California, with disease-specific differences in incidence by sex and race/ethnicity but not by urban versus suburban/rural county of residence. We report novel findings of a higher incidence of malignant PHEO in non-Hispanic Black Californians and a lower incidence of malignant PGL in Hispanic and Asian/Pacific Islander Californians. We also confirm higher incidence of ACC in women and non-Hispanic White Californians.

**Table 1. Age-adjusted incidence rate ratios (IRR) for ACC and malignant PHEO and PGL in California from 1992-2016 by sex, race/ethnicity, and urban versus suburban/rural county of residence. \*statistically significant versus reference population.**

Subgroup	ACC IRR (95% CI)	PHEO IRR (95% CI)	PGL IRR (95% CI)
Female	1.00 (reference)	1.00 (reference)	1.00 (reference)
Male	0.80 (0.69-0.92)*	1.24 (0.97-1.60)	1.49 (1.13-1.98)*
Non-Hispanic White	1.00 (reference)	1.00 (reference)	1.00 (reference)
Non-Hispanic Black	0.58 (0.40-0.82)*	1.65 (1.05-2.51)*	1.10 (0.62-1.82)
Hispanic	0.65 (0.54-0.78)*	0.93 (0.66-1.30)	0.60 (0.41-0.87)*
Asian/Pacific Islander	0.60 (0.46-0.76)*	0.92 (0.60-1.38)	0.60 (0.35-0.97)*
Rural/Suburban County	1.00 (reference)	1.00 (reference)	1.00 (reference)
Urban County	1.06 (0.91-1.24)	1.18 (0.89-1.60)	0.93 (0.69-1.27)