Unilateral (URA) and bilateral (BRA) renal agenesis are serious birth defects that can cause significant morbidity and mortality. Previous epidemiologic studies to evaluate the birth prevalence and influence of environmental and genetic factors on these birth defects have been hampered by small sample sizes and homogenous populations. This study readdressed the epidemiology of unilateral and bilateral renal agenesis using the Texas Birth Defects Registry from 1999-2004. Using Poisson regression we calculated associations between maternal and infant characteristics. In agreement with previous studies, we found an increased rate of unilateral renal agenesis in pre-term deliveries (OR 4.073, 95% CI 3.334-4.976) and diabetic mothers (OR 2.678, 95% CI 1.906-3.763). A decreased rate of URA was seen in the public health region encompassing Houston (OR 0.740, 95% CI 0.572-0.959). An increased rate of BRA was seen in mothers of parity of 4+ (OR 1.856, 95% CI 1.096-3.144) and births occurring in some public health regions along the Mexico border (OR 1.569, 95% CI 1.001-2.458). A decreased rate of BRA was seen in mothers under the age of 20 years (OR 0.552, 95% CI 0.318-0.960), which has been suggested in previous studies. Past research has suggested that black maternal race, older maternal age, and maternal education less than 12 years are associated with renal agenesis; however, our larger study did not support these findings. Implications of these findings are further discussed.