Chemistry 412 Seminar
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Wilms' Tumor: Childhood Cancer of the Kidney

Wilms tumor is one of the most common solid tumors found in children. In fact, 95% of solid kidney tumors found in children are Wilms' tumor. The average age at which this tumor is found is 36 months, with a range from 2 months-14 yrs. Development of the National Wilms' Tumor Study Group (NWTSG), in 1969, has helped to increase the survival rate of children affected with Wilms' tumor from 65% in 1970 to 90% in 1990 by studying the epidemiology, biology, and successful treatments of Wilms' patients. Over the years scientists have found that Wilms' tumor appears to be the result of sporadic and/or hereditary germ-line mutations found in one or more of several genes. Five different types of mutations have been found in Wilms' tumor: large deletions of part of the gene, nonsense or frameshift mutations affecting amino acids in the zinc fingers critical for DNA binding, missense mutations affecting the putative activation or repression domains, and mutations affecting preventing correct soliciting. These mutations affect critical cellular pathways during tumorigenesis and development. At the present time, mutations in the Wilms' Tumor gene-1 (WT1), located on the short arm of chromosome 11, have been found in 10-20% of the Wilms' tumors that have been studied. The WT1 gene may function as either an activator or repressor of gene expression and the protein may work at either the transcriptional or post-transcriptional level. The affects of mutations in both the WT2 gene and the b-catenin pathway are also being studied. This presentation will look at the NWTSG, the three mutations currently being studied, and the diagnosis, staging and treatment of Wilms' tumor.

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