## RENAL TUMORS

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Childhood renal tumors are caused by cancer cells growing in the kidneys. There are two kidneys in the human body filtering the blood, removing waste, making urine and maintaining our body's fluid balance by preserving essentials vitamins and regulating some of our body's hormonal systems.

The most common childhood kidney tumor is nephroblastoma, or Wilms tumor, which is also the sixth most common childhood malignancy with about 500 cases diagnosed annually in the United States.[1,2] In the older adolescent, the most common renal tumor is renal cell carcinoma.[1] Additional less common types of childhood renal tumors include rhabdoid tumors, clear cell sarcoma, congenital mesoblastic nephroma, and Ewing sarcoma of the kidney.

Renal Tumors make up around seven percent of all childhood cancers and ten percent of all malignancies in younger children ages one to four. [1-3] The clinical presentation of renal tumors is similar among all subtypes and typically includes an abdominal mass, blood in the urine, high blood pressure, and sometimes abdominal pain. [1,3]

Wilms tumor can affect either one or both kidneys, which is referred to as bilateral Wilms tumor and is present in 5-10% of cases. [1] To understand the full extent of the disease, labs as well as imaging studies.

The stage of Wilms tumor is based on its location and extent of disease ranging from stage I- stage V. Tumor solely confined to a single kidney is considered stage I. Stage II is any extension of the tumor into the renal vessels or renal capsule. Stage III is when there is any remaining disease in the abdomen after resection of the tumor or if there was biopsy was done prior to tumor resection.[1] Distant metastasis typically to the lung, bone or brain (or spread) is stage IV, and if the disease is in both kidneys it is considered stage V.[1] Wilms tumor is diagnosed based on a tissue sample with either a complete resection of the affected kidney or less commonly a biopsy.

Wilms tumor can be associated with different congenital anomalies or conditions in up to 15% of cases and is more often the case in patients with bilateral Wilms. [1-3] The most common congenital abnormalities include aniridia, or partial or complete loss of the colored portion of the eyes, genitourinary abnormalities, and hemihypertrophy (one side of body larger than the other). [1,3] The associated genetic syndromes are WAGR syndrome, Denys-Drash syndrome, Perlman Syndrome, Fanconi Anemia, and Beckwith-Wiedemann. [1,3] Patients with these syndromes are screened for Wilms tumor with abdominal ultrasonography every 3 months until they reach at least age 8 years.

A pediatric multidisciplinary team including pediatric oncologists, radiation oncologist, and surgeons will consider several factors to determine the appropriate therapy. These factors include the stage of the disease, histopathology, and the genetics of the tumor. Histopathology refers to how the tumor appears microscopically and can be considered either favorable histology or unfavorable histology with anaplasia. The genetics of tumor evaluate the chromosomal alterations of the tumor; currently the most prognostically important gain of chromosome 1q, and loss of heterozygosity of 1p and 16q.[1,3] The usual treatment of Wilms tumor will involve surgery and chemotherapy. Few low risk patients only require surgery. In more advanced stage disease, radiation therapy is also needed and is very effective in treating Wilms tumor.

Renal Cell carcinoma (RCC) is the second most common pediatric renal malignancy representing 5% of all childhood renal tumors.[1,3] RCC typically occurs in older adolescents ages 15-19 and is uncommon in children younger than 15. In pediatrics, RCC is often associated with the chromosomal translocation, TFE3. [1,3] Renal Cell carcinoma is associated with the inherited conditions von Hippel Lindau and Tuberous Sclerosis. [1,3] The prognosis of RCC is based on the stage

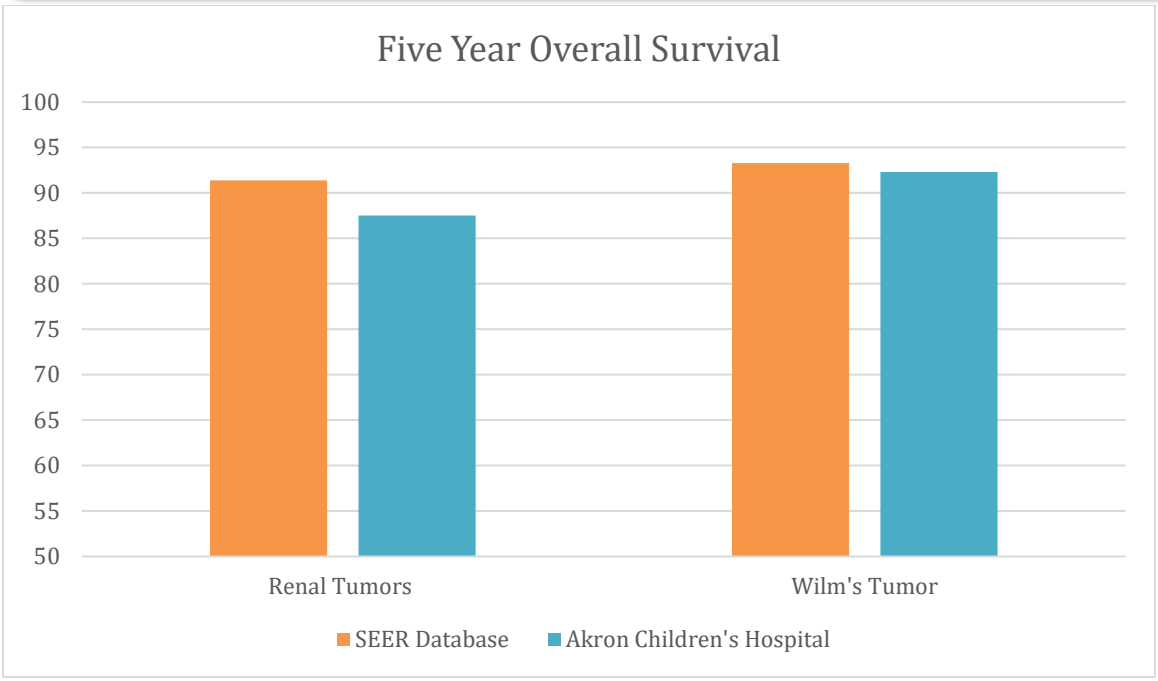
(I-IV) and local lymph node involvement. Surgery is the main stay of therapy. Renal cell carcinomas are often not responsive to chemotherapy and more targeted therapies including vascular endothelial growth factor inhibitors are being used. [1,3] The primary prognostic factor for RCC is stage of disease.

Clear Cell carcinoma represent 3-5% of pediatric renal tumors and has a 2:1 male predominance. [1,3] Unfortunately, clear cell carcinoma can be difficult to treat and has the potential of metastasizing to lung, bone, brain and soft tissue. This is a rare tumor and consideration should be taken to enrolling on a clinical trial and with a multidisciplinary team. Typically, the treatment involves surgery, radiation and chemotherapy.

Rhabdoid tumor of the kidney represents 2% of pediatric renal tumors and presents in infants and young children. [1,3] These tumors are very aggressive and respond poorly to both chemotherapy and radiotherapy and have a high degree of recurrence. Rhabdoid tumors often present in an advanced stage and these tumors commonly have a SMARCb1 chromosome mutation. [1,3] The prognosis of rhabdoid tumors is based on age at diagnosis, stage of the disease and whether they have lesions in the brain (CNS tumors). Rhabdoid tumors are often treated on clinical trials due to the aggressive and rare nature of this disease.

We reviewed our experience at Akron Children's Hospital treating patients with renal tumors from January 2011 to December 2016. During this time, we diagnosed 16 new cases of Renal Tumors with an average of 2.5 cases annually (1 to 5 cases per year). At Akron Children's Hospital we have an average of 90-100 new diagnoses a year, so this accounts for around 2-3% of our newly diagnosed malignancies. A majority, 81%, were Wilms tumor, but we also saw two cases of Renal Cell carcinoma and one case of Clear Cell carcinoma. Our Renal Cell carcinoma patients at Akron Children's Hospital were in the older adolescent group ages 15-17, and Wilms tumor occurred in our patients from ages 10 month to 6 years of age. At Akron Children's we had male predominance during this time of 1.6:1 for Wilms tumor, which is typically reported epidemiologically as 0.92:1 male to female radio.[1] Of our Wilms tumor patients, 85% were Caucasian, 15% were African American.

In reviewing the National Cancer Institute, surveillance, epidemiology and end results program database (SEER) the overall incidence from 2011-2017 of Renal Tumors in children 0-19 was 7.1%.[4] The overall survival at five years of our patients at Akron Children's Hospital with Renal Tumors was 87.5%, which is minimally lower than the national SEER database that report a 5-year relative survival of 91.4%.[4] This difference is likely due to the high proportion of renal cell carcinoma that we have had during the 2011-2016 time frame which represented 12.5% of our renal tumors. Our five-year overall survival during this time for Wilms tumor patients was 92.3%, while the relative five-year relative survival in the SEER data was 93.2%. [4]




Citations: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4) Browse the SEER cancer Statistics Review 1975-2018. (n.d.). Retrieved May 05, 2021, fromhttps://seer.cancer.gov/csr/1975_2018/browse_csr.php?sectionSEL=2&pageSEL=sect_02_table.06