Clear-cell sarcoma of the kidney (CCSK) is a rare pediatric renal tumor representing one of the unfavorable histologies in the National Wilms Tumor Study Group (NWTSG). CCSK is associated with a wider distribution of metastases, the most common site being the bones. Other metastatic sites reported include lung, abdomen, retroperitoneum, brain, and liver. Approximately 20% of documented CCSK metastases occurred at least 3 years after diagnosis; some occurred as long as 10 years later. CCSK frequently affects children aged 2 to 3 years. The mean age of presentation is 36 months. Incidence of CCSK is less common in children older than 3 years. The cause of CCSK is unknown and appears to be unrelated to Wilms tumor (WT). There are no known accepted specific chromosomal translocations associated with CCSK.

Clinical Signs and Symptoms:

The clinical signs and symptoms of CCSK are very similar to WT. As in WT, patients present with an abdominal mass that was incidentally noted during a routine examination, or by a caregiver while caring for the child.

- Asymptomatic abdominal mass, usually unilateral, unicentric
- Abdominal pain
- Malaise
- Hematuria
- Fever
- Hypertension- in 25% of patients
- Subscapular hemorrhage with rapid abdominal enlargement, anemia, and hypertension
- Metastatic disease in lungs, liver, and regional lymph nodes
- Bone pain

Diagnostic Workup:

- Complete history of the illness including pain (location, duration, and intensity); presence of abdominal mass; hematuria
Renal Tumors - Clear-cell Sarcoma of the Kidney

- Physical exam (unilateral fixed abdominal mass)
- Lab studies: CBC, urinalysis, renal, and liver function tests
- Abdominal ultrasound to determine tumor size and shape, vessel involvement, presence of thrombus in the inferior vena cava and right atrium
- Abdominal CT to assist with planning of surgery
- Chest radiograph to assess for metastases
- Chest CT to find lesions not noted on a chest X ray
- Biopsy and staging to determine histology and (A -2) stage of the disease (Hockenberry-Eaton, 1998)
- Bone scan and skeletal survey

**Histology:**

CCSK has nine major (A –3) histologic patterns: classic, myxoid, sclerosing, cellular, epithelioid, palisading, spindle, storiform, and anaplastic; virtually all tumors contain multiple patterns that blend together. The classic histologic pattern of CCSK is characterized by cells arranged in cords, nests, or groups surrounded by thin fibrovascular septa. A moderate amount of clear intercellular matrix separates the cord cells, giving a clear appearance; hence the designation CCSK.

The clear cell appearance of the tumor is caused by loose spacing of the round or oval cord cells with intervening intercellular clear mucoid matrix. The classic pattern is predominant, and often blends in with the variant pattern and may be seen in cases of metastatic tumors. The other eight patterns are recognized as variant and usually blend smoothly with the classic pattern or with another variant. Hyperchromasia, nuclear gigantism and atypical mitosis characterize the anaplastic pattern. Anaplastic lesions also have been associated with an over-expression of p53 gene.

**Treatment:**

Patients in all stages of CCSK are treated following the regimen for Wilms tumor with diffuse anaplasia. At presentation, (A –4) radical nephrectomy is the initial treatment of choice if the lesion is respectable. This is followed by radiotherapy and chemotherapy. If there are questions regarding the size or extension of the lesion, a biopsy is performed and chemotherapy is administered, followed by surgical resection after a response has been obtained. Chemotherapy agents used include cyclophosphamide, etoposide, vincristine, and doxorubicin for 24 weeks. Doxorubicin has been clearly shown to improve outcome and survival in CCSK.

**Future Directions:**

Similar to WT, CCSK investigations focus on maximizing efficacy of chemotherapeutic agents while minimizing immediate and long term side-effects.
Helpful Web Links Clear Cell Sarcoma of the Kidney

**E-Medicine**
This website contains information related to clear cell sarcoma of the kidney and discussions on treatment modalities.
http://www.emedicine.com/ped/topic3018.htm

**Doctor’s doctor.com, Torrance, CA**
http://www.thedoctorsdoctor.com/diseases/kidney_clearcellsarcoma.htm

**Texas Children’s Hospital – A Review of Renal Neoplasms**
http://sup.ultrakohl.com/uscap/abs-1998/hicks98.htm

Related [www.Cure4kids.org](http://www.cure4kids.org) Seminars

**Seminar #379 Clear Cell Sarcoma of Kidney**
Presenter: Hiroto Inaba, MD, PhD, Christine Fuller, MD and Fred Laningham, MD
http://www.cure4kids.org/seminar/379
Appendix:

A – 1  Gross anatomy of clear cell sarcoma of the kidney

![Image of clear cell sarcoma of the kidney](image-url)

Courtesy of C. Fuller MD, St. Jude Children’s Research Hospital

A – 2  Staging of CCSK

<table>
<thead>
<tr>
<th>Stage</th>
<th>Criteria</th>
</tr>
</thead>
<tbody>
<tr>
<td>Stage I</td>
<td>Tumor is limited to the kidney and is completely resected; renal capsule is intact, no evidence of rupture. No involvement of the vessels of the renal sinus and no evidence of tumor at or beyond the margins of resection.</td>
</tr>
<tr>
<td>Stage II</td>
<td>Tumor extends beyond the kidney and is completely resected; regional extension is present, involvement of the blood vessels outside renal parenchyma (including renal sinus). Biopsy prior to surgery or spillage of tumor during surgery confined to the flank without involvement of the peritoneal surface. No evidence of tumor at or beyond the margins of resection.</td>
</tr>
<tr>
<td>Stage III</td>
<td>Presence of abdominal lymph node involvement (renal hilar, para-aortic and beyond). Tumor penetrates peritoneal surface. Tumor implants on peritoneal surface. Gross microscopic evidence of the tumor present after resection. Resection is incomplete because of vital structures involvement. Tumor spillage is not confined to the flank.</td>
</tr>
<tr>
<td>Stage IV</td>
<td>Hematogenous metastasis (lung, bones, liver, brain) Lymph node metastasis extends beyond the abdomino-pelvic region</td>
</tr>
<tr>
<td>Stage V</td>
<td>Bilateral renal involvement at diagnosis. Like WT, each side is staged individually using above criteria</td>
</tr>
</tbody>
</table>
A - 3 CCSK histologic pattern

- Myxoid pattern (50%)
- Sclerosing pattern (35%)
- Cellular pattern (26%)
- Epithelioid pattern (trabecular or acinar type) (13%)
- Palisading (Verocay body) pattern (11%)
- Spindle cell pattern (7%)
- Storiform pattern (4%)
- Anaplastic pattern (2.6%)

Classic Histology pattern

Spindle Pattern CCSK

Courtesy of C. Fuller, MD, St. Jude Children's Research Hospital
A – 4  Radical Nephrectomy – organs removed and postero-lateral incision

Hollywood Urology, Dr. Tom Shannon, Urologist, Australia
www.hollywoodurology.com/ radneph.html

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