



**Society of Radiologists in Ultrasound
2012 Toshiba Residents Program**

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Clinical History: 1 year old boy with a left abdominal mass noted on exam during well-child checkup. No past medical or surgical history. No recent fever, weight loss, anorexia, or hematuria.

Figures:

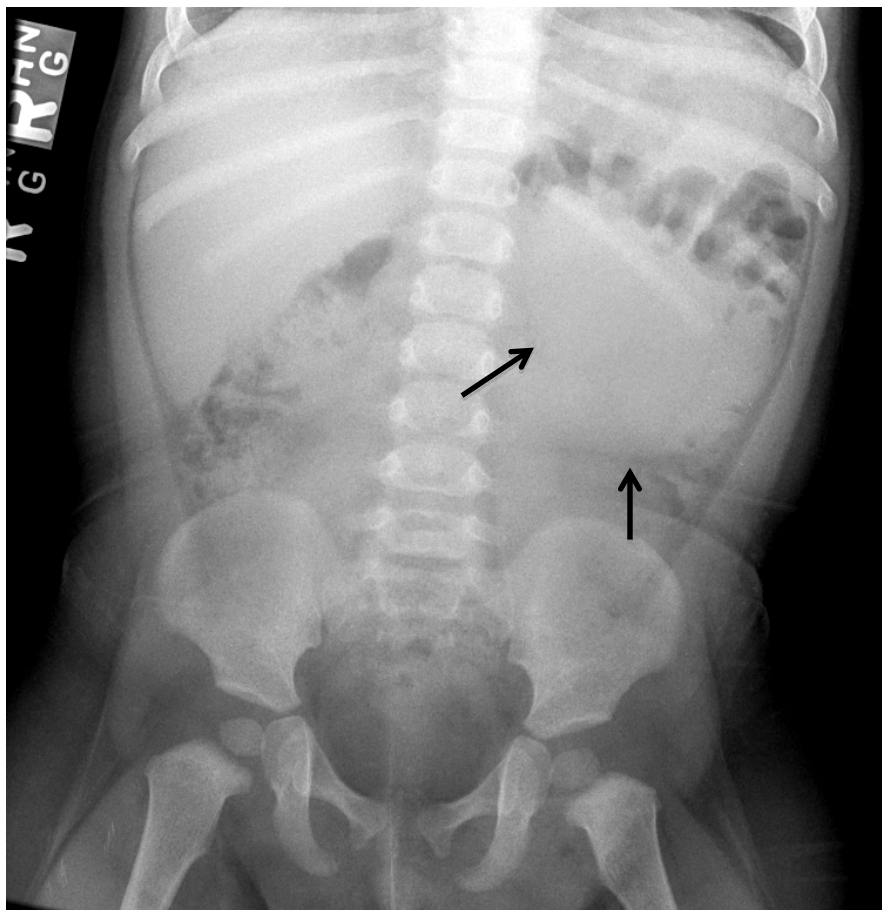


Figure 1: Frontal radiograph of the abdomen demonstrates soft tissue mass in the left abdomen (arrows), with displacement of air-filled bowel.

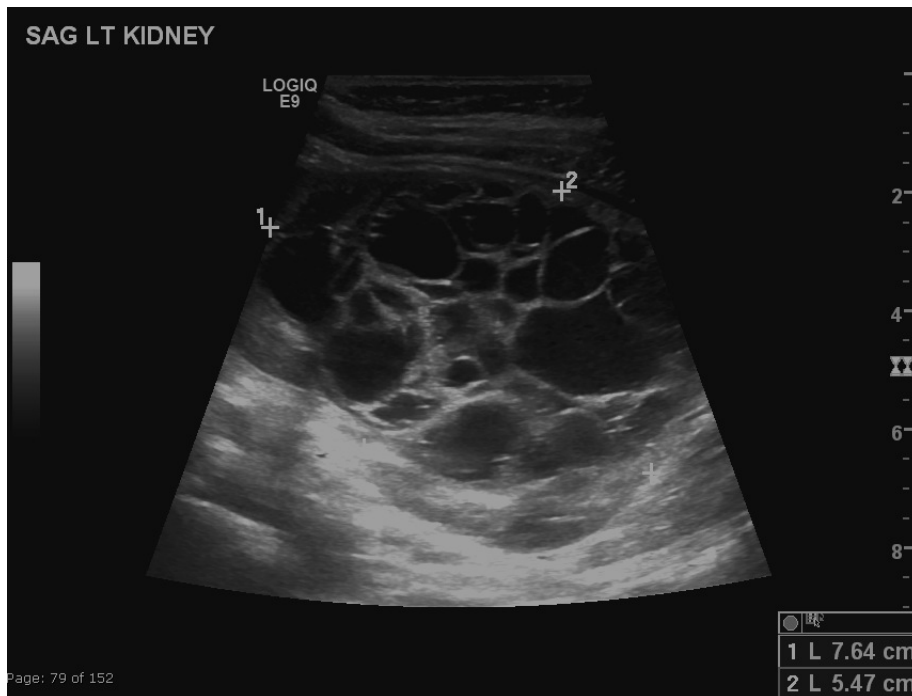


Figure 2a: Grey scale ultrasound in the sagittal plane through the left kidney demonstrates a cystic mass measuring 7.6cm in greatest dimension.

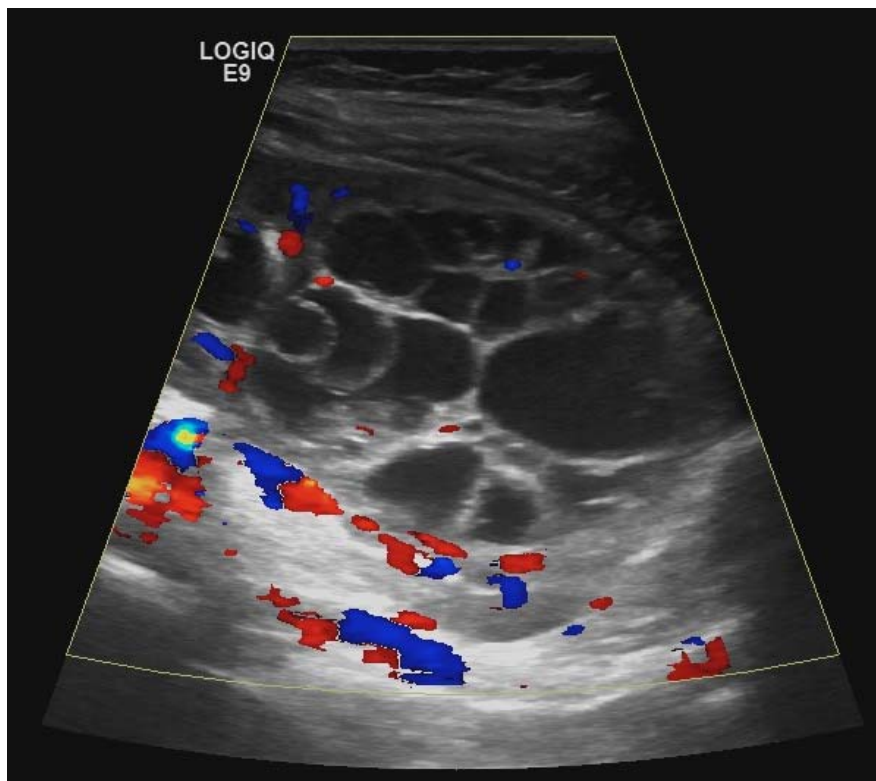


Figure 2b: Color Doppler of the left renal mass demonstrates flow in the renal parenchyma peripheral to the mass and within the intervening fibrous septa. No large soft tissue component.

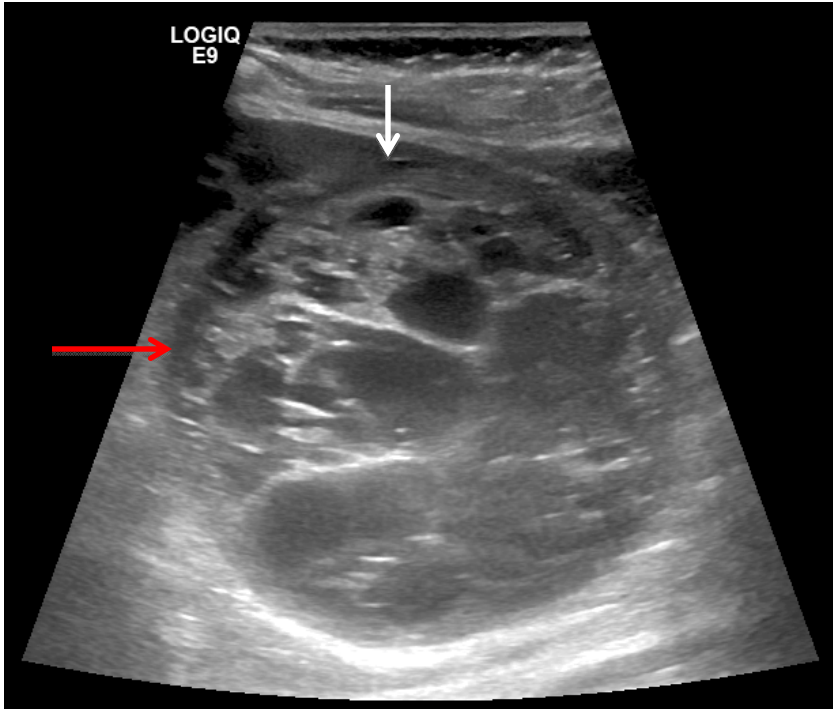


Figure 2c: Sagittal grey scale demonstrates the well circumscribed mass and normal renal parenchyma surrounding the mass (arrow) with a characteristic "claw sign," reflecting intra-renal location. There is a distinct border that suggests a capsule (red arrow).

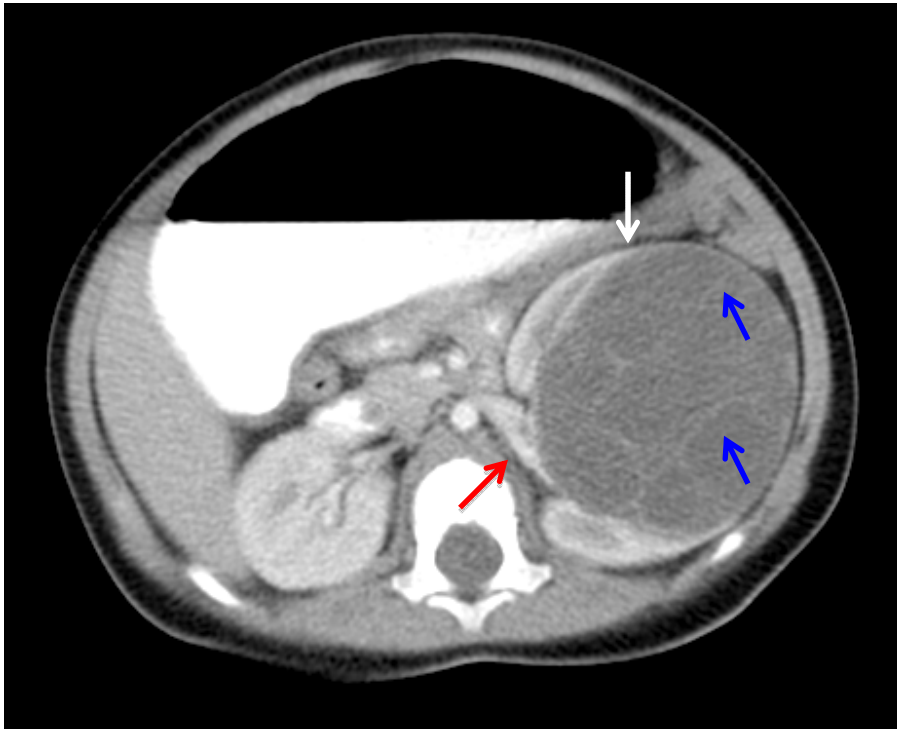


Figure 3a: Axial contrast-enhanced CT of the abdomen through the kidneys demonstrates a large cystic mass in the left kidney with enhancing septa (blue arrows). Again is noted the

renal parenchyma enveloping the mass (white arrow), “claw sign”. The partially visualized left renal vein homogeneously enhances (red arrow).



Figure 3b,c: Coronal and Sagittal reformatted images (respectively), demonstrate the left cystic renal mass.

Diagnosis:

Cystic Nephroma, pathology proven at nephrectomy.

Discussion:

Cystic nephroma is a rare, non-familial neoplasm that has been found in patients as young as three months of age into the seventies (rare before 3 months); the vast majority found to be between 3 and 24 months and in the fifth and sixth decades of life, demonstrating a biphasic age distribution.¹ There is a gender difference, with a greater percentage of males presenting before age 4; whereas the majority of patients older than 4 are female.¹ The most common presentation is an incidentally noted mass on abdominal exam.²

Multilocular cystic renal tumors are a group of benign neoplasms of which cystic nephroma and cystic partially differentiated nephroblastoma (CPDN) are distinct subgroups. These neoplasms are identical on imaging but differ histologically. Cystic nephromas are composed of cysts and septa, well-demarcated from the adjacent renal parenchyma, with flattened or cuboidal epithelium lining the cysts.³ CPDN, in addition, contain blastemal cells, and may or may not contain embryonal stromal or epithelial cells.³ Although both histologic subtypes are present in all age groups, CPDN is more common in the younger age group that is predominantly male.⁴

Radiographic findings are non-specific and may show a soft tissue mass in a retroperitoneal location with displacement of adjacent structures, depending on size.

Calcifications may be apparent on plain film, although uncommon, and would be present in the soft tissue components, septa or capsule, and will have a curvilinear orientation.¹ In the pediatric population, ultrasound is the first-line imaging modality and a multicystic intrarenal mass is the predominant feature. The variability in sonographic features depends on the amount of soft tissue components (fibrous septa and capsule) and size of cysts.¹ The smaller the cysts, the greater acoustic interfaces and the more mass-like will be the sonographic appearance. Given the intra-renal location, a common radiologic finding is the “claw sign,” related to the acute angles formed by normal renal parenchyma due to the mass arising from the kidney.

Cross-sectional imaging is commonly performed and cystic nephroma as well as CPDN appear as a well-marginated, multicystic mass with variable enhancement of the septa on CT. The more complex the cystic contents, the more mass-like will be the appearance on CT.¹ Extension into the renal pelvis can be visualized on CT and may account for a presentation of hematuria. Magnetic resonance imaging will demonstrate a multicystic mass, the contents of which are T2 hyperintense and variable intensity on T1 weighted sequences (related to the amount of protein or hemorrhage) surrounded by hypointense fibrous septa and capsule.⁵

The differential diagnosis of the multilocular cystic renal tumors in the pediatric population is: cystic Wilms tumor, renal cell carcinoma, clear cell sarcoma, mesoblastic nephroma, and the segmental variant of multicystic dysplastic kidney. The degree of hemorrhage and necrosis in the solid malignancies is what can mimic multilocular cystic renal tumors.⁶ The histologic diagnosis cannot be predicted by imaging alone, thus, complete resection is required for diagnosis and treatment.⁶ Complete excision with tumor-free margins is curative and if there is a recurrence, radiation and/or chemotherapy may be warranted.³ The amount of immature elements in CPDN may predict aggressive behavior,³ thus noninvasive monitoring is suggested in cases of cystic partially differentiated nephroblastoma.

References:

¹Madewell JE, Goldman SM, Davis CJ Jr, Hartman DS, Feigin DS, Lichtenstein JE. Multilocular cystic nephroma: a radiographic-pathologic correlation of 58 patients. *Radiology* 1983 146: 309-321.

²Banner MP, Pollack HM, Chatten J, Witzleben C. Multilocular renal cysts: radiologic-pathologic correlation. *AJR*. 1981 136: 239-247.

³Joshi VV, Beckwith JB. Multilocular cyst of the kidney (cystic nephroma) and cystic, partially differentiated nephroblastoma: terminology and criteria for diagnosis. *Cancer* 1989 64: 466-479.

⁴Lonergan GJ, Martinez-Leon MI, Agrons GA, Montemarano H, Suarez ES. Nephrogenic rests, nephroblastomatosis, and associated lesions of the kidney. *RadioGraphics* 1998 18: 947-968.

⁵Silver IA, Boag AH, Soboleski DA. Multilocular Cystic Renal Tumor: Cystic Nephroma *Radiographics*. 2008 28: 1221-1225.

⁶Agrons GA, Wagner BJ, Davidson AJ, Suarez ES. Multilocular cystic renal tumor in children: radiologic-pathologic correlation. *RadioGraphics*. 1995 15: 653-669.

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