The most common cancers overall in children are leukemia (28%), brain and spinal tumors (26%).
They are followed by tumors that mainly present as an abdominal tumor: Neuroblastoma (8%), Nephroblastoma (5%) and Lymphoma (8%) (ref).

Pediatric abdominal tumors are often very large at initial presentation, because most children come to attention because someone noted severe abdominal distention.

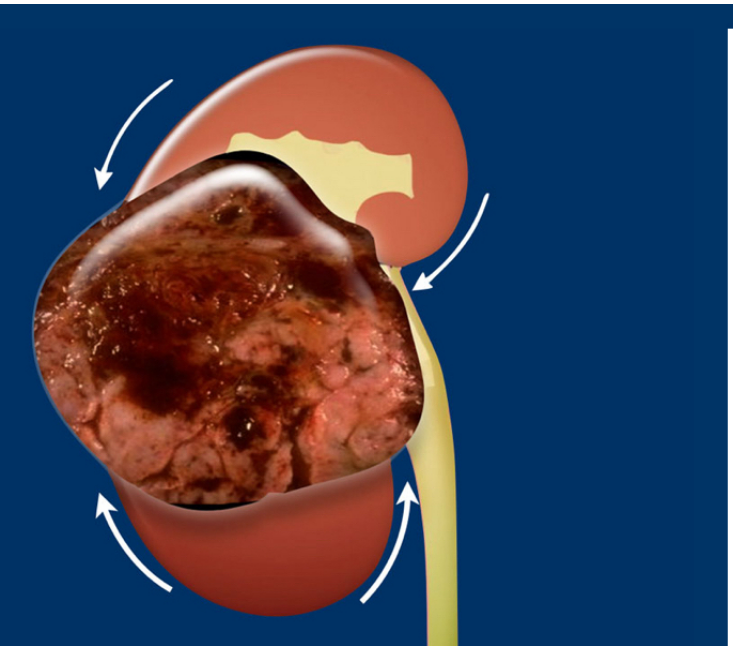
It may seem a contradiction, but in very large tumors, it is usually more difficult to ascertain the organ of origin.

The most common intra-abdominal tumors in children are:

- Neuroblastoma: 30% of all cases
- Nephroblastoma or Wilms' tumor: 25%
- Lymphomas:15%
- Germ cell tumors: 9%
- Hepatoblastoma: 9%
- Rhabdomyosarcoma: 4%
- Hepatocellular carcinoma: 1,5%

Key features of Abdominal tumors

Neuroblastoma	Malaise, pain and abdominal mass; opsoclonus myoclonus syndrome or metastatic disease. US inhomogeneous and echogenic, with bright calcifications. VMA and HVA levels in the urine
Nephroblastoma	Most common renal tumor in children Distended painless abdomen, sometimes hematuria or hypertension. US claw sign, small tumors will move synchronous with the kidney.
Hodgkin lymphoma	Cervical lymph node enlargement, and mediastinal masses. Rarely confined to the abdomen.
Non-Hodgkin	Para-aortic and mesenteric nodes. Spleen involvement.
Leukemia	May present with abdominal involvement, mostly kidneys.
Hepatoblastoma	Abdominal mass, otherwise asymptomatic Usually high AFP.
Hepatocellular carcinoma	Uncommon malignant liver tumor in teenagers. Abdominal mass, jaundice, pain, AFP high.
Germ cell tumor	Usually in testis or ovary but may present as abdominal tumor. High AFP, beta-HCG and CA-125.



In renal tumors the “claw sign” is often present.
This is seen when a part of the kidney is draped around the tumor like a claw (figure).

Before we discuss the different abdominal tumors, it is good to realize that the definitive diagnosis is usually made by pathologic tissue examination.

Most tumors are biopsied before treatment.
However renal tumors in children between six months and nine years are not biopsied because the likelihood of it being a nephroblastoma is so high that the risk of a wrong diagnosis outweighs the risk of tumor spill during a biopsy, especially in diffuse anaplastic nephroblastoma.

Neuroblastoma

Features of Neuroblastoma

- Embryonic tumors originating from the sympatho-adrenal lineage of the neural crest.
- 50% arises in adrenal gland.
- Most common cancer in babies.
- Third-most common in children (90% < 5 years).
- Main differential diagnosis of nephroblastoma.
- Clinical: Pain - abdominal mass - raccoon eyes - opsoclonus myoclonus syndrome
- High levels of Vanillyl Mandelic Acid and HomoVanillic Acid in urine.
- Diagnosis is based on biopsy.

Neuroblastomas are embryonic tumors originating from the sympatho-adrenal lineage of the neural crest.
About half of the tumors arise from the adrenal glands.
Other sites of origin are the thoracic and lumbal paravertebral sympathic chain.
A minority develops in the neck.

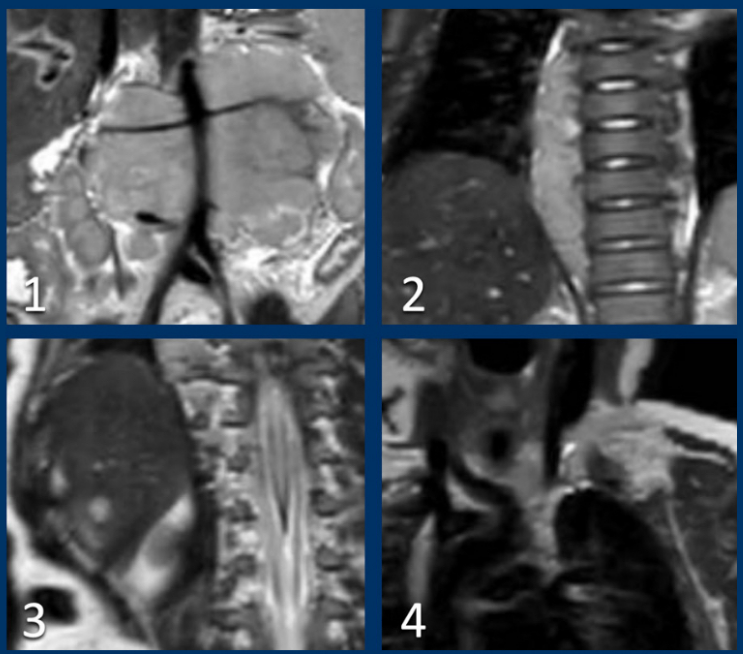
The clinical presentation is variable. Common complaints are pain and an abdominal mass. Neurologic signs due to intraspinal extension can occur. A typical manifestation is “raccoon eyes”, which is periorbital ecchymosis due to metastatic infiltration of the orbital area.

A minority of the patients present with opsoclonus myoclonus syndrome, a neurological disorder characterized by rapid, multi-directional eye movements (opsoclonus), quick, involuntary muscle jerks (myoclonus), uncoordinated movement (ataxia), irritability, and sleep disturbance.

The prognosis depends on the stage of the tumor.
For a low grade tumor the 5-year survival is > 90%.
For high risk tumors (stage 4 and tumors with MYCN amplification) it is around 50%.

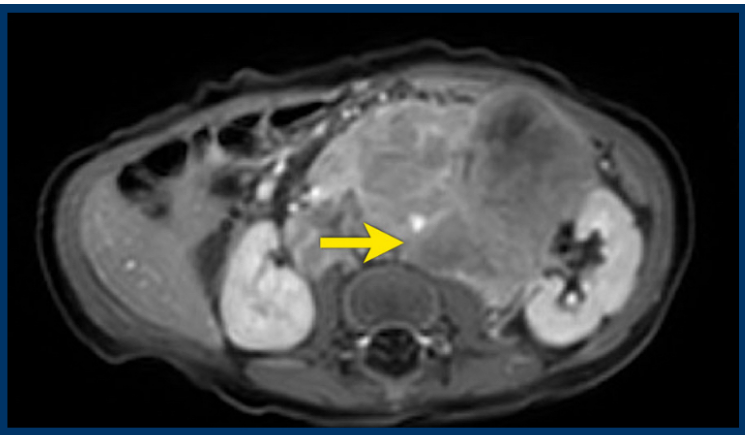
Imaging findings in Neuroblastoma

- Encasement of the splanchnic vessels
- Calcifications
- Intraspinal extension
- Lymph node involvement.
- Thrombosis of the renal vein as a propagation of an adrenal vein thrombosis.
- Invasion of the kidney or liver.
- Distant metastasis in the liver and bones, especially the vertebral column, the pelvis, and the skull.
- Additional brain imaging is guided by the results of MIBG scintigraphy.



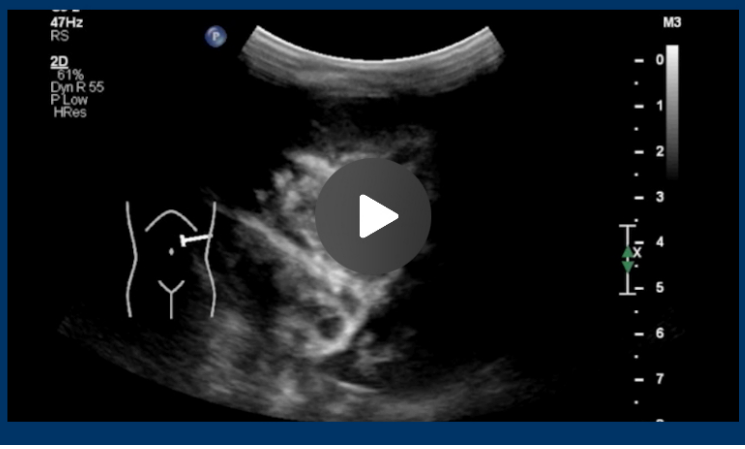
The findings are:

1. Mass with encasement of the aorta and splanchnic vessels.
2. Extension along the thoracic vertebral columns but no intraspinal invasion.
3. Small liver metastases.
4. Left supraclavicular mass.



Same patient.

This is an axial gadolinium enhanced T1W-image with fat suppression.
It shows the encasement of the vessels.
Notice the tumor extension posterior to the aorta, which is displaced away from the vertebral column (arrow).



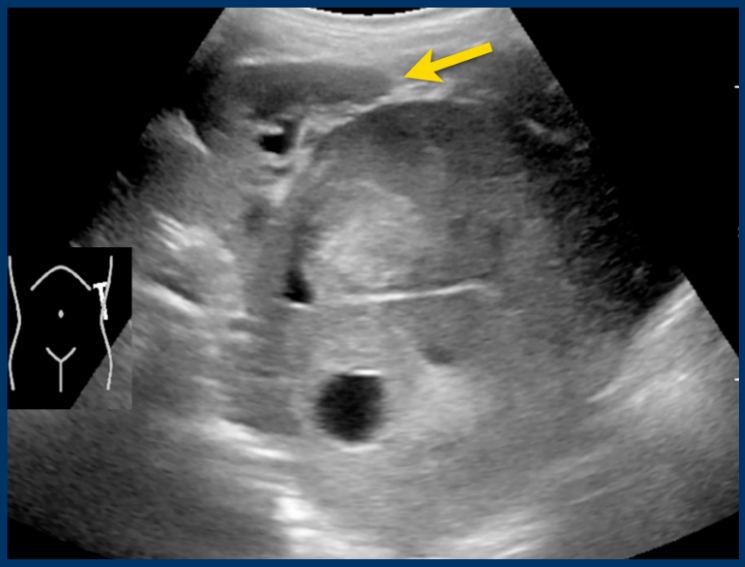
Ultrasound of a fifteen-month-old boy, who was first suspected of having a tumor in the left kidney.

Ultrasound shows a mass adjacent to the medial upper pole of the left kidney. It seems to be separate from the kidney. The mass is very inhomogeneous and has multiple calcifications.

These findings are more compatible with a neuroblastoma than a nephroblastoma.

Renal tumors

Renal tumors in children will be discussed in more detail in a separate article.
Here we only show some common findings.



Nephroblastomas

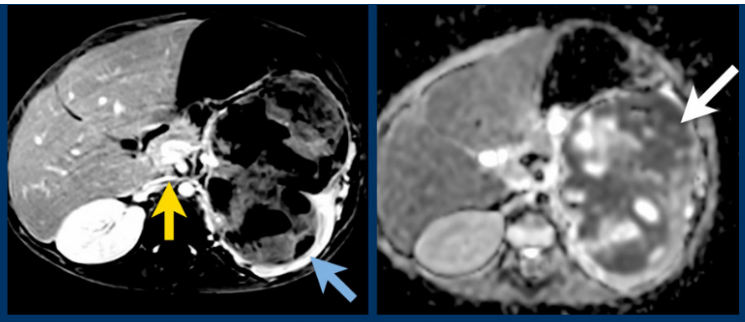
More than 90% of renal tumors in children are nephroblastomas - also called Wilms tumor.

The peak age is 2 - 3 years.

Frequently these tumors present with a distended painless abdomen and they are often very large at presentation. Sometimes they present with hematuria, abdominal pain or hypertension.

Smaller tumors are found during sonographic screening in children with syndromes which predispose to nephroblastoma, like Beckwith-Wiedemann syndrome and Denys-Drash syndrome (see the webpage on renal tumors in children).

Bilateral nephroblastomas are often syndrome related. The lungs are the most frequent site of metastases. Liver and bone metastases are rare.

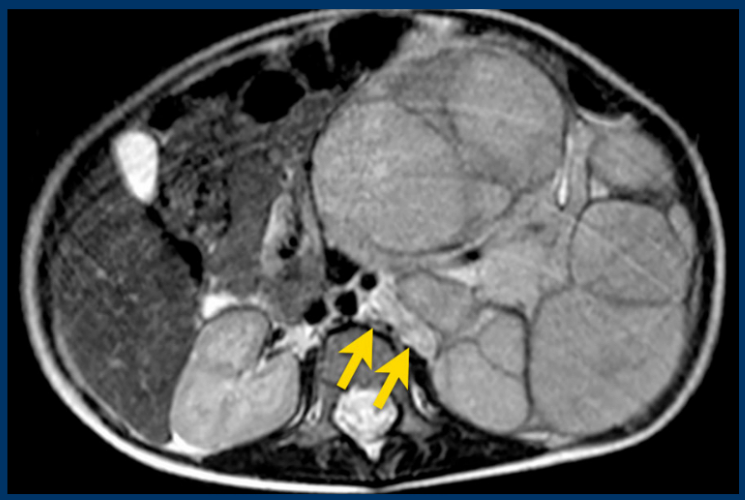


A. The tumor enhances less than the peripheral remnant of normal renal tissue (blue arrow). The left renal vein is open (yellow arrow). Solid parts of the tumor show diffusion restriction (white arrow)

Ultrasound
The initial imaging is usually done by ultrasound. Smaller tumors will be seen to move synchronous with the kidney. Large tumors will not move.

As mentioned before, it is often possible to detect a remnant of the kidney draped around the tumor, the claw sign. The remnant can have a dilated calyx due to obstruction of the pelvis.

Small tumors are usually homogenic and echogenic.
Larger tumors are more inhomogeneous with cystic or necrotic parts and hemorrhage.
10% of the nephroblastomas have fine calcifications.

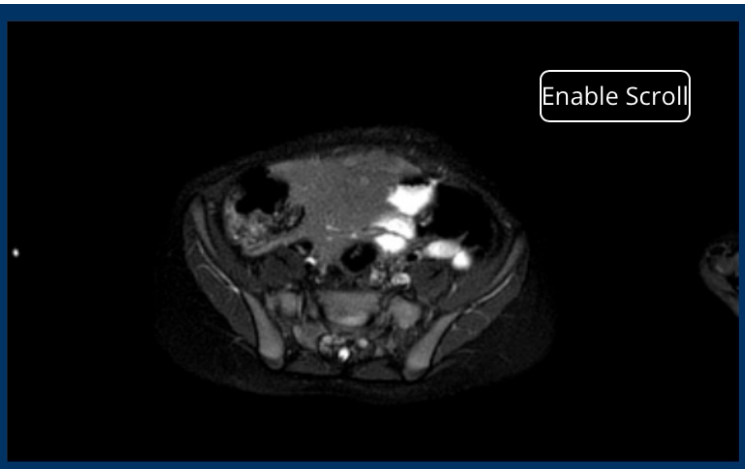


Left sided nephroblastoma in a two-year-old girl. Note the para-aortal lymph node metastasis (arrow).

Once you are sure of the renal origin of the tumor, study the other kidney for tumor or nephroblastomatosis (see below).
Search for enlarged para-aortal lymph nodes.
Search with color Doppler for patency of the renal vein and of the inferior caval vein, as a nephroblastoma tends to grow into the renal vein and inferior caval vein.

Also image the liver for metastases, although these are rare in nephroblastoma.
The finding of a liver metastasis should urge you to look for an alternative diagnosis like a rhabdoid tumor.

A CT chest is performed for the assessment of pulmonary metastases.



MRI
The next imaging step is a MRI of the abdomen.

Nephroblastomas are mostly inhomogeneous, with decreased signal intensity on T1 and increased signal intensity on T2. Necrotic cystic parts are often present. Gadolinium enhancement is inhomogeneous and less than the enhancement of normal renal parenchyma.

Solid parts of the tumor will show restricted diffusion. Hemorrhage is often present. Hemorrhagic areas will also show restricted diffusion, so look on the T1-images for signs of bleeding.
Sometimes a disruption of the tumor capsule is seen. Intraperitoneal rupture is a more severe complication than retroperitoneal rupture.

MRI can nicely demonstrate tumor thrombus in the renal vein and inferior caval vein, and lymph node enlargement. It allows accurate and repeatable measurements of the tumor on the initial and follow-up examinations.

Classification of Nephroblastoma

Stage 1	Tumor limited to the kidney, completely resected
Stage 2	Tumor not limited to the kidney, but completely resected
Stage 3	Tumor not completely resected, confined to the abdomen. Includes peroperative tumor rupture
Stage 4	Metastatic spread to lungs, liver, brain, or bone
Stage 5	Bilateral tumor

Classification of Nephroblastoma

The classification of nephroblastomas is done after resection of the kidney.

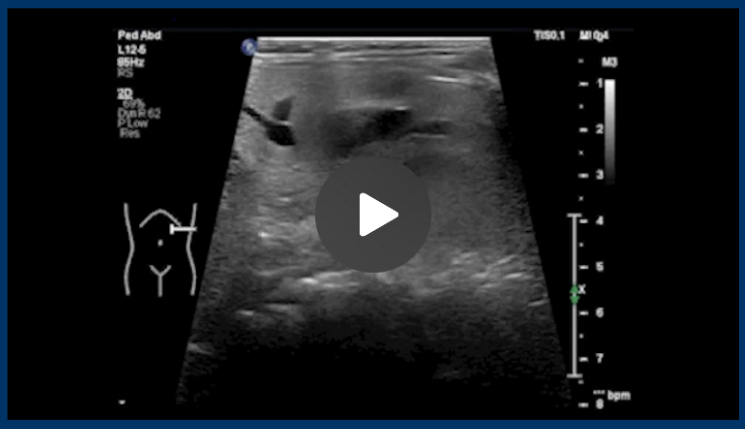
Other Renal tumors in Children

Nephroblastomatosis
Malignant rhabdoid tumor of the Kidney (MRTK)
Clear cell sarcoma of the Kidney (CCSK)
Congenital mesoblastic nephroma
Renal cell carcinoma (RCC)
Angiomyolipoma

Other Renal tumors

More renal tumors will be discussed in a separate article.

Liver tumors

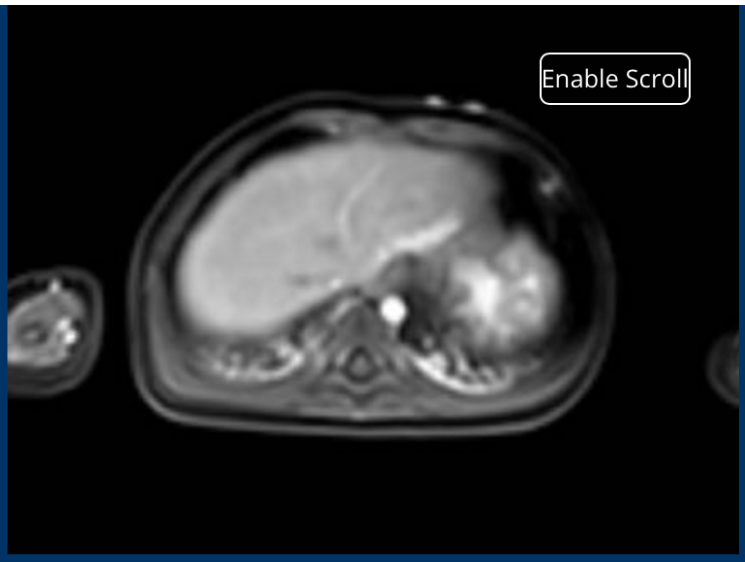


Hemangioendothelioma

Hemangioendothelioma of the liver is also known as infantile hemangioendothelioma or infantile hepatic hemangioma. It is a highly vascular tumor. These tumors can be solitary, multiple or diffuse.

Most are discovered as an abdominal mass in the first six months of life. They can lead to congestive heart failure and rarely to Kasabach-Merritt syndrome, a rare disease, in which the vascular tumor leads to decreased platelet counts and bleeding disorder.
AFP levels are mostly normal.

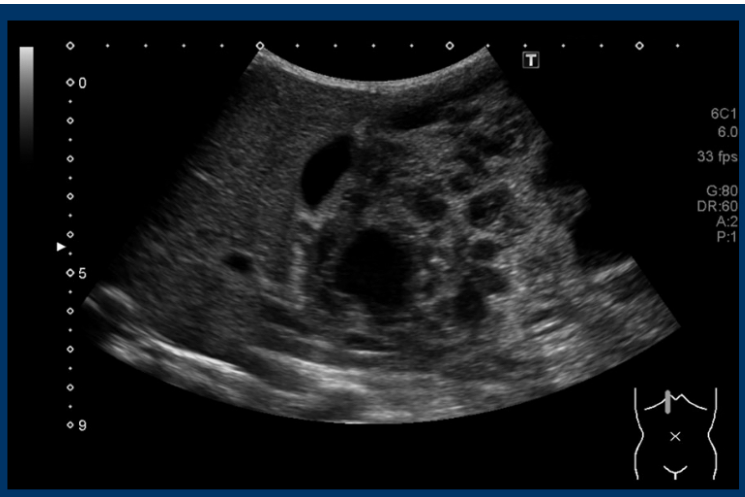
Ultrasound
On ultrasound a well-perfused tumor is seen. It can be hypoechoic or of mixed echogenicity. Unlike adult hepatic hemangiomas they are not echogenic. Calcifications are common.
Large arteries and veins are seen and the aorta may be wider than normal due to the large demand of the tumor and tapers distal to the celiac axis.



CT
On unenhanced CT calcifications are present in approximately half of the patients. After intravenous contrast the tumor shows peripheral enhancement with gradual filling-in. In larger tumors the center may not enhance at all.

MRI
On MRI a hemangioendothelioma has generally low signal intensity on T1 and high signal intensity on T2. After contrast the same filling-in is seen as on CT.
Most tumors will show spontaneous involution, and the prognosis is good.

Images
Scroll through the images. It is the same tumor as on the ultrasound.



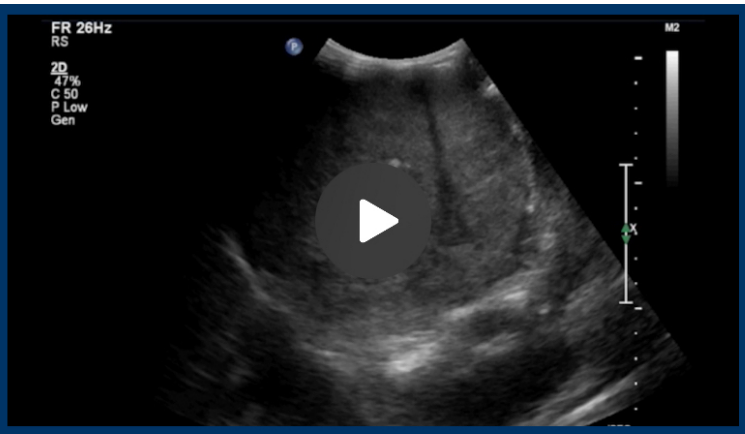
Mesenchymal hamartoma

Mesenchymal hamartomas are usually multicystic liver lesions, although they can rarely be solid. They are often large at presentation. Serum AFP levels are normal.

Ultrasound will show a multicystic lesion. MRI will demonstrate this as well. After Gadolinium some stromal enhancement can be seen.

Example 1
The image is of a two-year-old boy, who presented with a painless swelling of the abdomen. Ultrasound shows a large multicystic mass at the caudal side of the liver.

Continue with the MRI.



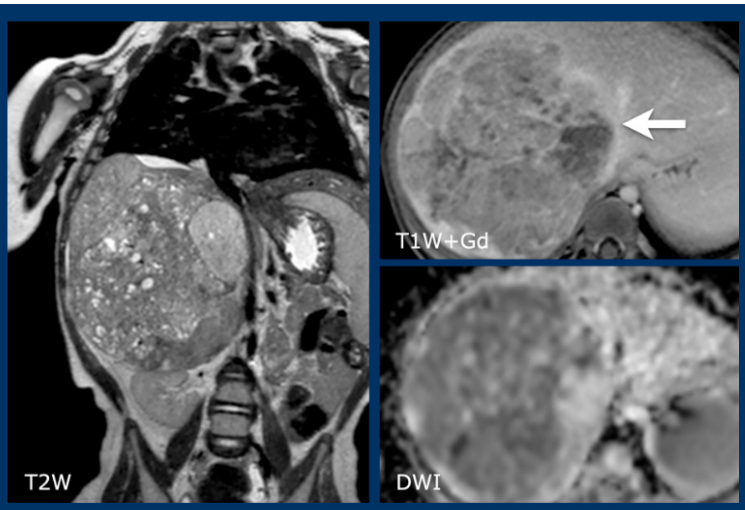
Hepatoblastoma

Hepatoblastoma is the most common malignant liver tumor in young children, while hepatocellular carcinoma presents in older children, mostly in their teens. [7]. Hepatoblastoma usually presents with an enlarged abdomen.

Ultrasound will generally show a well demarcated tumor. In larger tumors necrotic cysts and calcifications can be seen.

CT angiography is done preoperatively to define the relation between the tumor and the hepatic vessels.

MRI will better delineate the tumor. A hepatoblastoma has low signal intenstity on T1 and mixed signal intensity on T2. After Gadolinium patchy enhancement is seen.

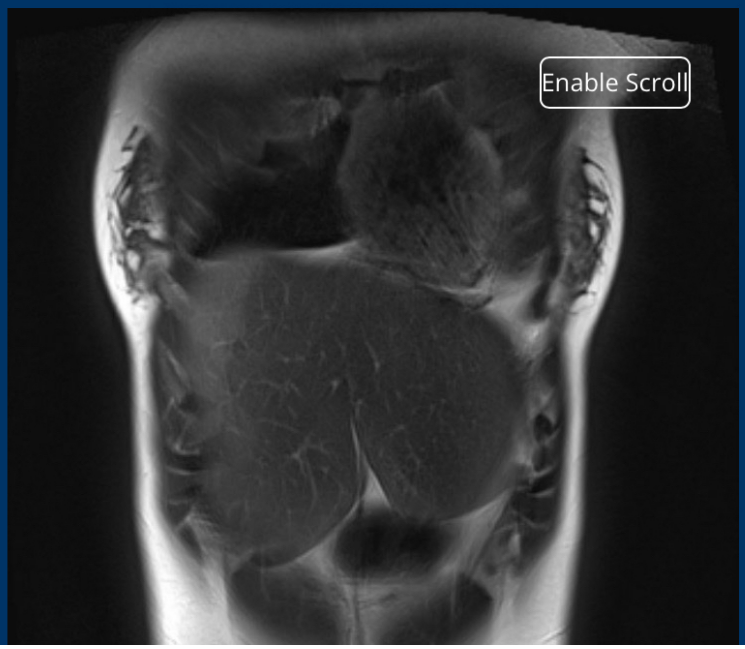


MRI shows a solid hepatic mass with multiple small cysts.

After contrast injection the mass is seen to be limited to the right posterior and anterior section, bordered by the middle hepatic vein (arrow).

The mass has moderate diffusion restriction.

Biopsy was compatible with an epithelial hepatoblastoma.



Hepatocellular carcinoma

This disease is rare in young children but can be seen in older children, mostly >10 years of age, although it has even been reported in children of five years of age. Underlying disease which predispose to HCC include hepatitis B and Tyrosinemia.

Tyrosinemia is a genetic disorder characterized by the failure to break down tyrosine, a building block of most proteins. Tyrosine and its byproducts will build up in organs and can lead to liver and kidney failure and an increased risk for HCC.

The tumor presents with abdominal mass, pain, or jaundice. AFP levels are elevated (although usually less elevated compared to AFP levels in hepatoblastoma).

Hodgkin and Non-Hodgkin

Hodgkin versus Non-Hodgkin

	Hodgkin disease	Non-Hodgkin
Cervical lymph nodes	**80%**	**<50%**
Para-aortic lymph nodes	35%	50%
Mesenteric lymph nodes	**5%**	**50%**
Mediastinum	**75%**	**25%**
Lung parenchyma	12%	5%
Liver	3%	15%
Spleen	**15%**	**>40%**
Gastro-intestinal tract	-	+
Kidney	-	+

There are two main types of lymphoma: Hodgkin lymphoma and non-Hodgkin lymphoma.

Hodgkin lymphoma more commonly manifests with cervical lymph node enlargement and mediastinal masses, while it is rarely confined to the abdomen.

Non-Hodgkin is more commonly located in the para-aortic and mesenteric lymph nodes and the spleen (table). Non-Hodgkin lymphoma presents more frequently with extra nodal disease than Hodgkin lymphoma.

For staging of Hodgkin lymphoma the Lugano classification is used [9], and for non-Hodgkin lymphoma the International Pediatric NHL staging system [10].

Ultrasound

On ultrasound enlarged lymph nodes are very hypoechoic. The almost anechoic aspect of the tumor is typical of malignant lymphoma. If the bowel is affected the layering of the bowel wall is lost.

MRI

On MRI masses are seen with some enhancement after Gadolinium and remarkable strong diffusion restriction. Another tumor that can show this marked diffusion restriction is a neuroblastoma, however these tumors are often much more heterogeneous with areas of necrosis and hemorrhage

PET-CT

18-F-FDG PET-CT is used for staging.

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Leukemia

Leukemia is the most common malignancy in children. It can present with abdominal involvement.

Leukemia can affect all solid abdominal organs.
The organs can be diffusely infiltrated or have a more nodular pattern.

The kidneys are affected in almost half of the patients with later stages of acute lymphoblastic leukemia. It can be uni- or bilateral, and there can be focal lesions or diffuse infiltration. The last has a rather typical appearance with a striated pattern around the calices, like in malignant lymphoma.

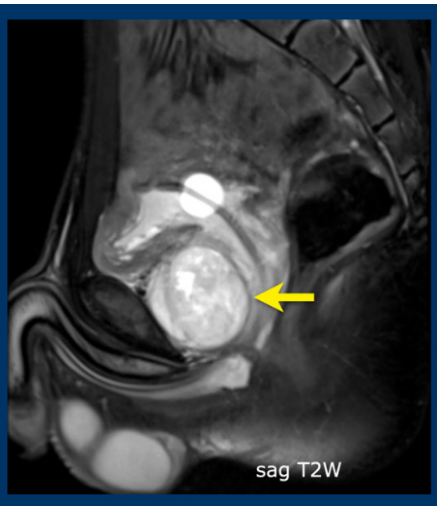
Rhabdomyosarcoma

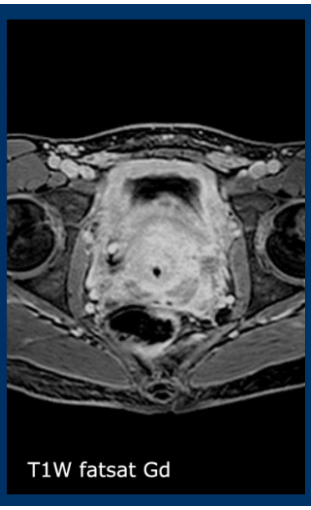
Rhabdomyosarcomas (RMS) are the most common soft tissue tumors in children and can develop almost anywhere but mostly in the head and neck region, including the orbit and in the genitourinary tract.

About 25% of all RMS arise in the lower abdomen, generally originating from the bladder, prostate or vagina, but they can arise almost anywhere, for instance along the biliary tract (where no striped muscle is present!).

The most common pathologic subtype is embryonal RMS, followed by alveolar RMS. The alveolar type has a worse prognosis.

The age of the patient, generally below 15 years and the location of the tumor in the prostate, bladder or vagina will point towards the diagnosis, while the imaging features are non-specific.





MRI

A sagittal image shows a tumor anterior to the bladder neck.

There is patchy enhancement.
DWI showed strong diffusion restriction (not shown).

The location of the tumor makes a rhabdomyosarcoma the most likely diagnosis.