Evaluating Common Intra-Abdominal Masses in Children — A Systematic Roentgenographic Approach

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An intra-abdominal mass in an infant or young child can be cancerous, and should be carefully and immediately evaluated. However, through a concern for thoroughness, numerous studies may be ordered, some of which are both physically and emotionally traumatic to the child and his family. These more complicated procedures are frequently unnecessary and often yield information that could have been obtained by the selective use of less traumatic techniques.

This article presents a systematic approach to the roentgenographic evaluation of neonates and children less than eight years old with intra-abdominal masses, especially those involving the urinary system and adrenal gland. By following a step-wise method—each new step related to findings obtained in earlier studies—most of the more complicated and invasive procedures can be avoided.

Abdominal masses in pediatric patients commonly involve the kidneys and bladder; adrenal lesions are less common. Other tumors such as lymphoma, teratoma, lymphangioma, rhabdomyosarcoma or lipoma are rarely found in the retroperitoneal space in children. Lesions of the liver, bile duct, spleen, pancreas and gastrointestinal tract are not discussed in this paper.

Once a mass has been palpated, abdominal roentgenography can pinpoint its location to the right or left upper quadrant, the retroperitoneal space, the gastrointestinal tract or mesentery. Often, several views including supine, prone, oblique and upright are considered necessary.

When the abdominal roentgenograms fail to adequately define the structures, other non-invasive diagnostic techniques are indicated.

- *Excretory pyelography* outlines intrarenal areas and demonstrates the effect of the mass on the urinary tract system.
- *Total body opacification*[1] occurs within the first 60 seconds of the excretory pyelogram. Solid structures with normal or increased vascularity are opaque; cystic structures are radiolucent. This procedure, therefore, can predict the probable solid or cystic nature of a mass. However, it is important to note that abscess, necrosis or hemorrhage within a solid tumor may also cause radiolucent areas.
- *Ultrasonography*[2] records sound reflections at tissue interfaces. Normal physiologic function is not necessary

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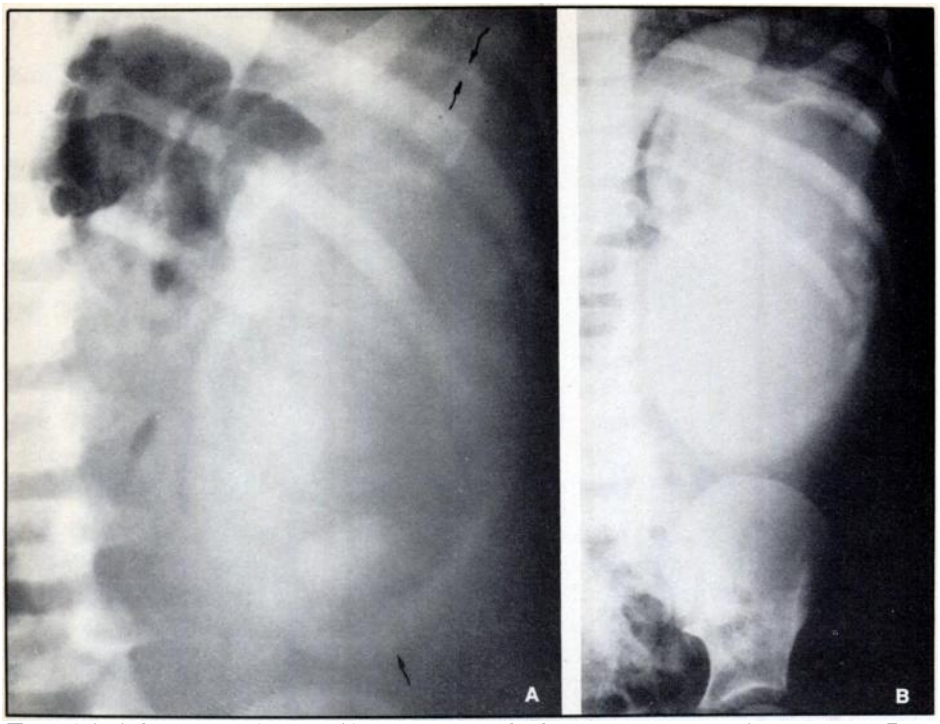


Fig. 1 (A). A four-year-old male with acute onset of left upper quadrant pain and mass. Early phase of the excretory pyelogram shows compressed renal tissue (arrows) due to hydronephrosis (crescent sign). (B.) Late phase of excretory pyelogram demonstrates collection of contrast material within the dilated renal system, the result of pelvi-ureteric obstruction.

to obtain these recordings. The density of the tissue and its elasticity, as well as the orientation of the ultrasound beam to tissue, must be evaluated. With B-mode ultrasonography, cystic structures appear as echo-free areas (anechoic) and solid structures as dots (echoic). Low-gain setting of the ultrasonogram indicates the size of the mass; high-gain setting indicates if it is solid.

- *Radioisotope scans* of the kidneys demonstrate the blood flow to a renal mass that was not visualized on excretory pyelography. Imaging with an isotope such as ^{99m}Tc glucohepatonate can differentiate renal tumors from pseudotumors. This modality is also useful in detecting incomplete rotation of the kidney.

A physical examination and clinical history, as well as the selective use of abdominal roentgenography, excretory pyelography, ultrasonography and isotope scanning usually establish the diagnosis. Only rarely, when the disease process complicates interpretation of these studies, are invasive methods, such as angiography, necessary.

Complete evaluation and a minimum number of procedures: these are the goals of the following systematic approach to diagnosing intra-abdominal masses in children. It should be remembered that cancers are rare, and that most tumors in children are benign.

RENAL MASSES

The most common abdominal mass in children is renal in origin. Excretory pyelography with early phase total body opacification provide findings within

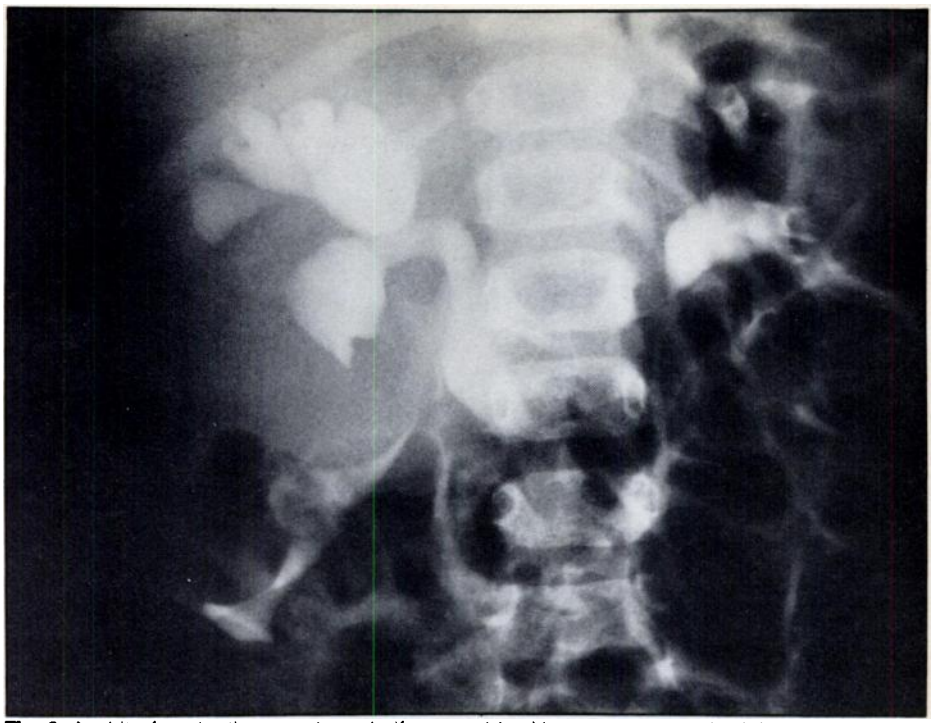


Fig. 2. A white female, three and one-half years old, with an asymptomatic right mass and mild hypertension. Excretory pyelogram demonstrates splaying of the calyceal system with partial obstruction by the mass, causing mild to moderate hydronephrosis.

three broad categories that distinguish various types of renal abnormalities.

These include:

1. *Calyceal distortion* from hydronephrosis, cystic kidney disease, abscess, benign and malignant renal tumors, as well as infiltrative diseases such as lymphomas and leukemia;

2. *Non-visualization* caused by total obstruction of the urinary tract, renal vein thrombosis or replacement of the kidney by infiltrating tumor;

3. *Duplication* of part of the urinary system.

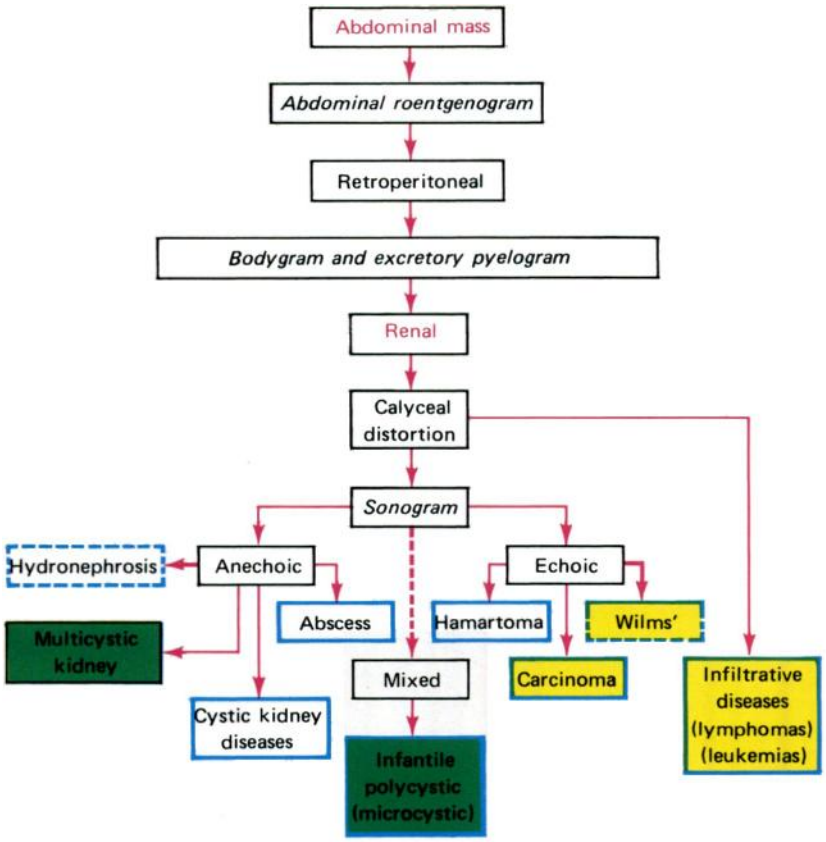
Calyceal Distortion

(FLOW CHART I)

Hydronephrosis (pyelocaliectasis) resulting from pelvi-ureteric obstruction is a common anomaly occurring at all ages in childhood. In most patients, the excretory pyelogram shows a dilated renal pelvis and calyces. With severe obstruction, the early finding on pyelography may be a rim of functioning tissue (crescent sign),[3] due to compression of the renal cortex by dilated calyces; on delayed roentgenograms, the contrast material can be seen within the dilated renal system. (Fig. 1.) A tumor of the pelviureteric tissue is extremely rare in children, but when it occurs, its extrinsic nature can usually be detected.

Wilms' tumor, exceedingly rare in neonates, is the most common solid renal tumor in children one year of age and older. Peak incidence occurs at about three years of age; the tumor is uncommon after eight years of age. The most striking feature of the disease is displacement and distortion of the pyelocalyceal system on excretory pyelography. (Fig. 2.) Total body opacification is unreliable, since lucent areas may rep-

FLOW CHART I



KEY FOR ALL FOLLOWING FIGURES

Neonate

Beyond neonate

Any age, common

Any age, moderately common

Any age, uncommon

Uncommon

Common (greater incidence)

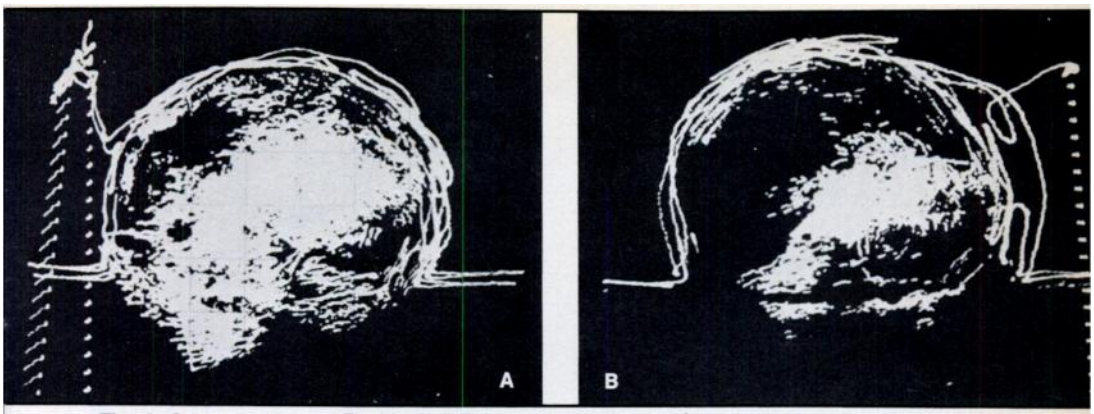


Fig. 3. Same patient as Fig. 2. (A.) This sonogram with high-gain setting is echo producing, indicating a solid mass. (B.) The low-gain sonogram outlines a large right renal mass and a normal left kidney. The combined findings of the excretory pyelogram and the sonogram indicate a Wilms' tumor.

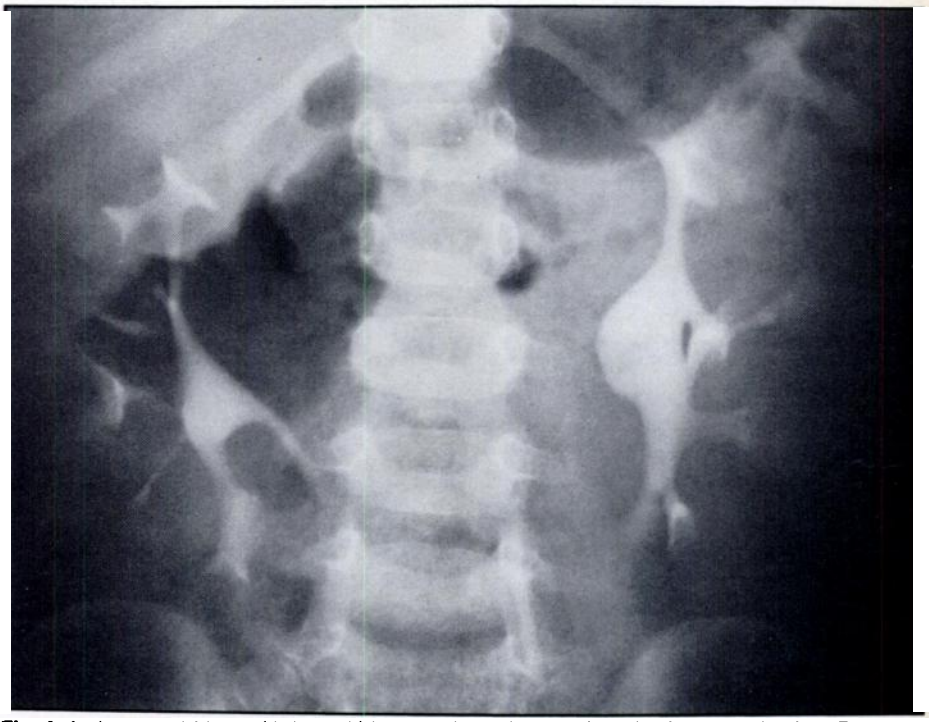


Fig. 4. A six-year-old boy with large kidneys palpated on routine physical examination. Excretory urogram shows an even pattern of marked stretching of the intrarenal structures. No obstruction of any portion of the calyceal structures is present. The patient had an anterior mediastinal mass and unilateral pleural effusion. Biopsy revealed lymphosarcoma.

resent hemorrhage and necrosis in a solid tumor, rather than a cystic lesion.[4]

The ultrasonogram of patients with Wilms' tumor demonstrates echoes, indicating a solid mass. (Fig. 3.) Hemorrhage and necrosis, which are often present, produce a mixed pattern of echoes and echo-free areas. Since blood

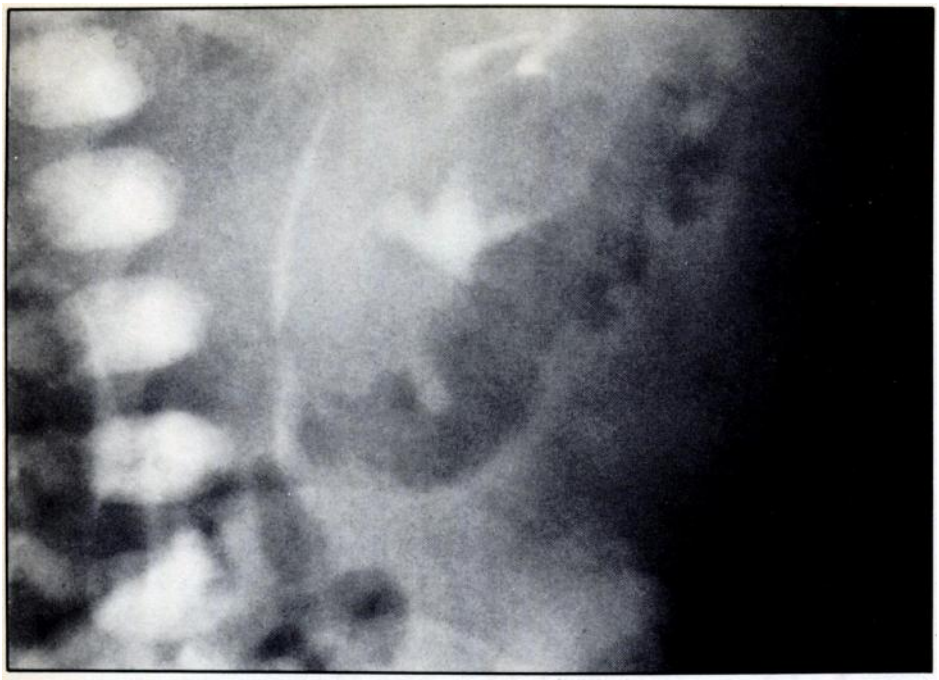


Fig. 5. Newborn with left renal mass demonstrating distortion and splaying of the pyelocalyceal system on excretory pyelography. Surgical specimen showed a congenital mesoblastic nephroma. (Courtesy of Dr. Walter Formen.)

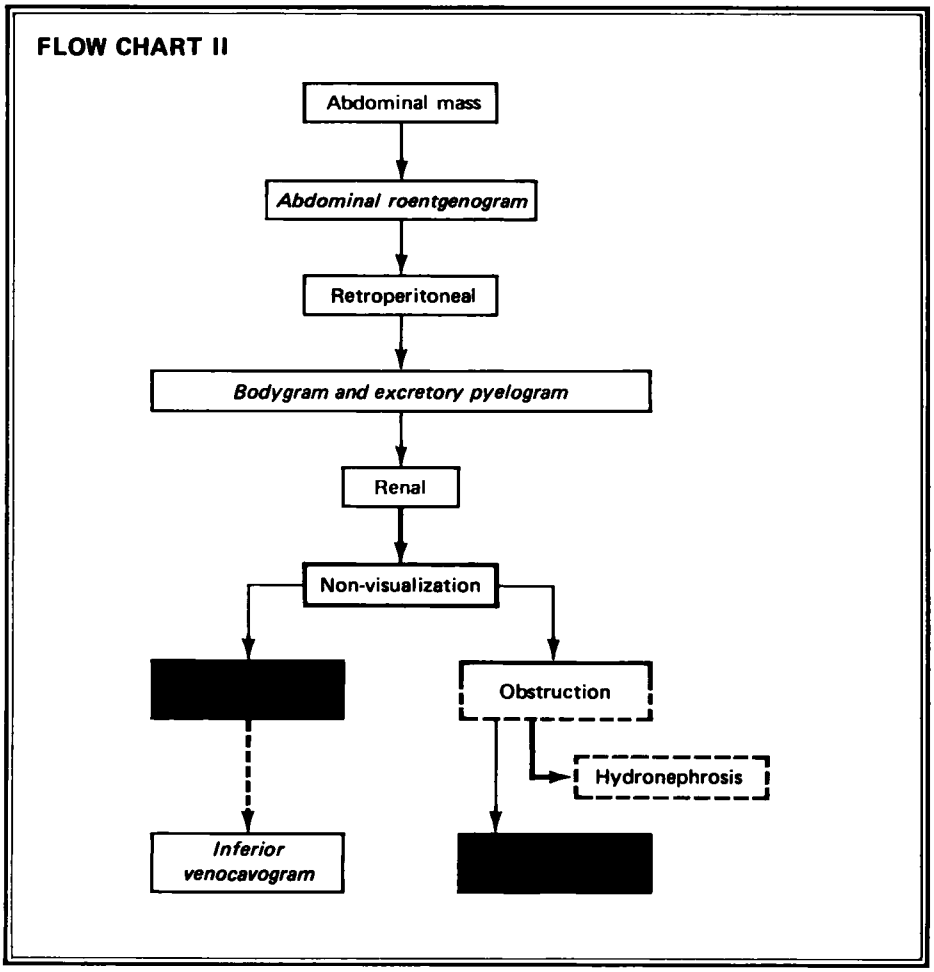
supply to Wilms' tumor is by the renal artery, or the renal capsular or inferior adrenal arteries that branch off the renal artery, anomalous blood supply is not a problem. Arteriography does not aid in the staging or management of a child with Wilms' tumor.[5] Angiography is not recommended by the "National Wilms' Tumor Study-1 Protocol."

Intrarenal splaying of the calyceal system by a cyst can occasionally be confused with Wilms' tumor on pyelography. Ultrasonography shows an anechoic pattern, supporting the diagnosis of a cyst.

A renal cell carcinoma cannot be differentiated from Wilms' tumor roentgenographically. However, the patient's age may provide a clue; carcinoma of the kidney, although uncommon in children, tends to occur beyond the Wilms' tumor age group. Calcification, which may develop in either condition, is more common in those patients with renal cell carcinomas.

An enlarged kidney, unilateral or bilateral, may be due to infiltrative diseases including the lymphomas, especially lymphosarcoma, or leukemia. The calyces appear to be stretched out on excretory pyelography, mimicking renal edema associated with the nephrotic syndrome. (Fig. 4.) Further evaluation includes careful palpation for lymphadenopathy and chest X-rays to rule out the presence of a mediastinal mass and pleural fluid. Bone marrow examination may be indicated. However, these procedures often obviate the need for renal arteriography and renal biopsy.

Most congenital renal tumors are benign hamartomas, commonly referred to as congenital mesoblastic nephroma,[6,7] fetal hamartoma[8] and mesenchymal renal neoplasms.[9] Congenital mesoblastic nephroma is generally discovered



in young patients during the first weeks of life as an asymptomatic abdominal mass.

Excretory pyelography demonstrates distortion of the pyelocalyceal system of the affected kidney similar to that seen in patients with Wilms' tumor. (Fig. 5.) Ultrasonography shows an echoic mass. Umbilical aortography often reveals neovascularity, which gives a false impression of malignancy; this procedure does not aid in management and carries a risk to the patient. Evaluation other than excretory pyelography prior to surgery is not necessary for these patients. Survival following nephrectomy is excellent.

Non-visualization

(FLOW CHART II)

Multicystic kidney, considered by many investigators to be a form of hydronephrosis,[11,12] is the most common renal mass in newborns. Cancer is rare in this age group. During the bodygram phase of the excretory pyelogram, the mass appears lucent, relative to adjacent organs with good blood supply. (Fig. 6.) The ultrasonogram is anechoic, confirming the cystic nature of this lesion.

Renal vein thrombosis[13,14] presents as an enlarged kidney, caused by renal engorgement and complicated by gross or

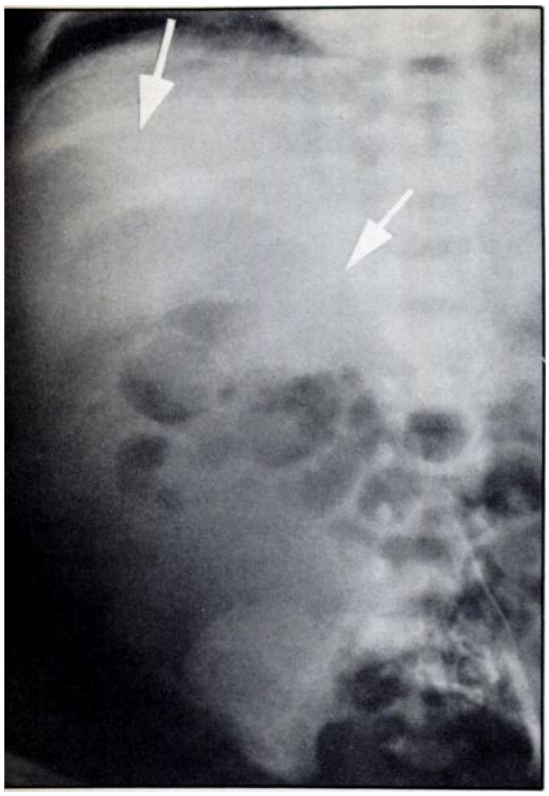


Fig. 6. The bodygram phase of the excretory pyelogram shows a lucent area (the upper border is indicated by arrows), relative to the visualized adjacent organs, indicating the cystic nature of the mass. Multicystic kidney was found at surgery.

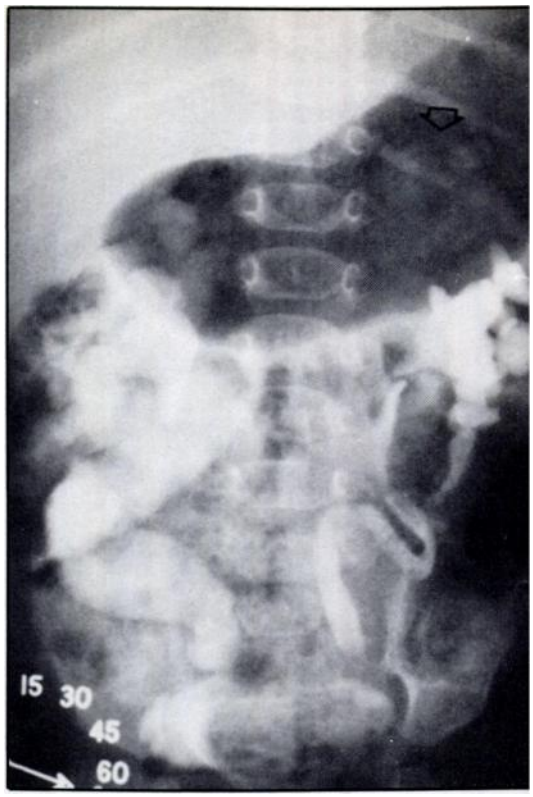


Fig. 7. Excretory pyelogram of a five-month-old infant with bilateral renal masses. The right kidney shows dilation of the duplex system. The upper pole of the left kidney is not visualized (arrow) due to severe hydronephrosis. The lower segment of the duplex system on the left is depressed downward by the hydronephrotic upper pole. The lucency within the bladder represents the ureterocele.

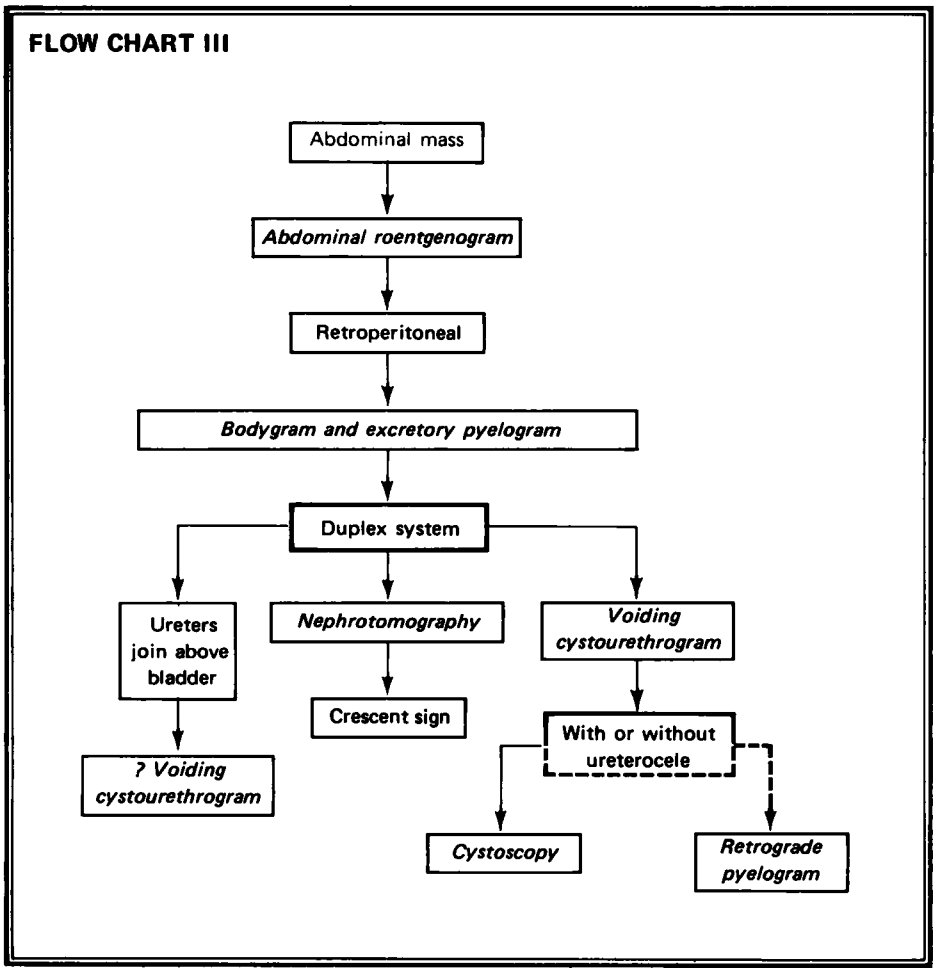
microscopic hematuria and an elevated blood pressure. Many children with renal vein thrombosis are born to mothers with diabetes. In some neonates and older infants, diarrhea or sepsis and severe dehydration are the first clinical indications of the disease. There is non-visualization of the affected kidney on excretory pyelography. Clinical and roentgenographic findings are usually sufficient for proper diagnosis. However, if in doubt and treatment depends on definitive diagnosis, an inferior venacavogram can be performed to determine whether the thrombus has extended from the renal vein to the vena cava.

Non-visualization of a kidney occurs in approximately 10 percent of patients with Wilms' tumor. This tumor can cause loss of renal function by its extension into the renal vein, obstruction of the renal pelvis or ureter or by replacement of renal tissue. Ultrasonography is very helpful in determining if an enlarged kidney represents a solid tumor, which in the pediatric age group is assumed to be Wilms' tumor.

Duplication

(FLOW CHART III)

Duplication of the kidney and ureter[15] often present with an enlarged kidney.



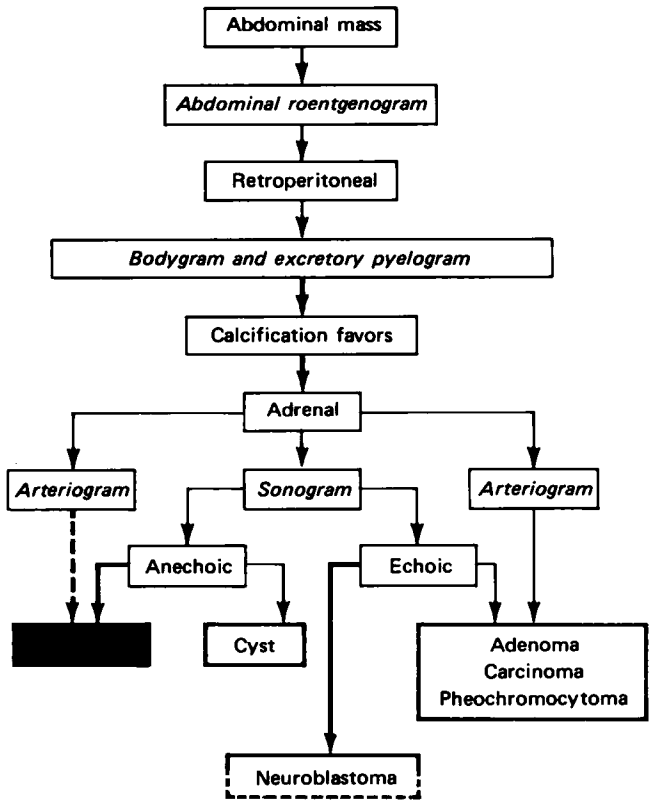
The ectopic ureter drains the upper pole of the kidney, frequently forming a ureterocele in the caudal portion of the bladder. The ureterocele then causes obstruction with subsequent hydronephrosis and kidney enlargement. Excretory pyelography may show a large kidney with little or no visualization of the upper pole; the lower pole calyces may be depressed by the hydronephrotic upper pole. (Fig. 7.) With an obstructed and dilated upper pole and ureter, the lower renal segment may be displaced laterally, mimicking a neuroblastoma. Contrast material often reveals a filling defect caused by the ureterocele.

ADRENAL MASSES

(FLOW CHART IV)

Enlargement of the adrenal gland is far less common than enlargement of the kidney. Overall, the most likely cause of an enlarged adrenal gland in a newborn infant is hemorrhage,[16,17] often associated with jaundice.[18] Adrenal insufficiency is occasionally evident, but only when the hemorrhage is bilateral. Neuroblastoma, which comprises approximately 10 percent of all childhood cancers, is another cause of adrenal enlargement. Adenoma, carcinoma and

FLOW CHART IV



pheochromocytoma of the adrenal gland may also be present.

It is sometimes difficult to diagnose the cause of an adrenal mass by initial roentgenographic studies. In patients with adrenal hemorrhage, total body opacification usually demonstrates a lucency, while in those with neuroblastoma, roentgenography shows a blush due to increased blood supply to the tumor. However, exceptions to the blush appearance have been reported. The pyelogram reveals downward displacement of the kidney in both adrenal hemorrhage and neuroblastoma. On sonography, an adrenal hemorrhage is anechoic; a neuroblastoma is usually echoic, although homogeneity of neuroblastoma tissue may occasionally produce an anechoic pattern.

However, distinctions between these two conditions become apparent over a short period of time. Since adrenal hemorrhage contracts rapidly, the orientation of the kidney begins to return to normal within days, and is completely normal in several weeks. In hemorrhage, calcification commonly occurs within a few days to two weeks, takes on a curvilinear shape and contracts rapidly as the hematoma shrinks. (Fig. 8.) In neuroblastoma, calcification is punc-

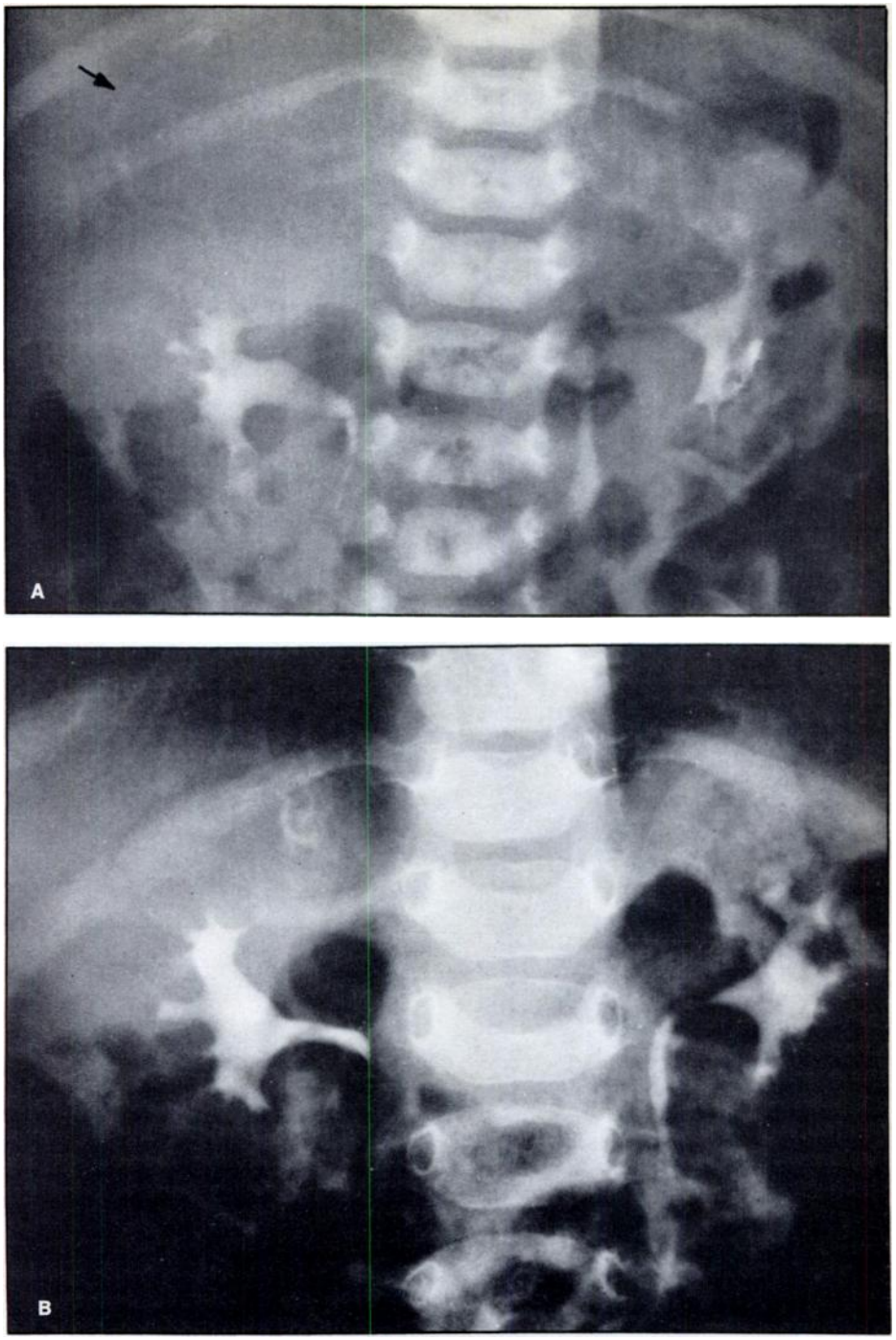
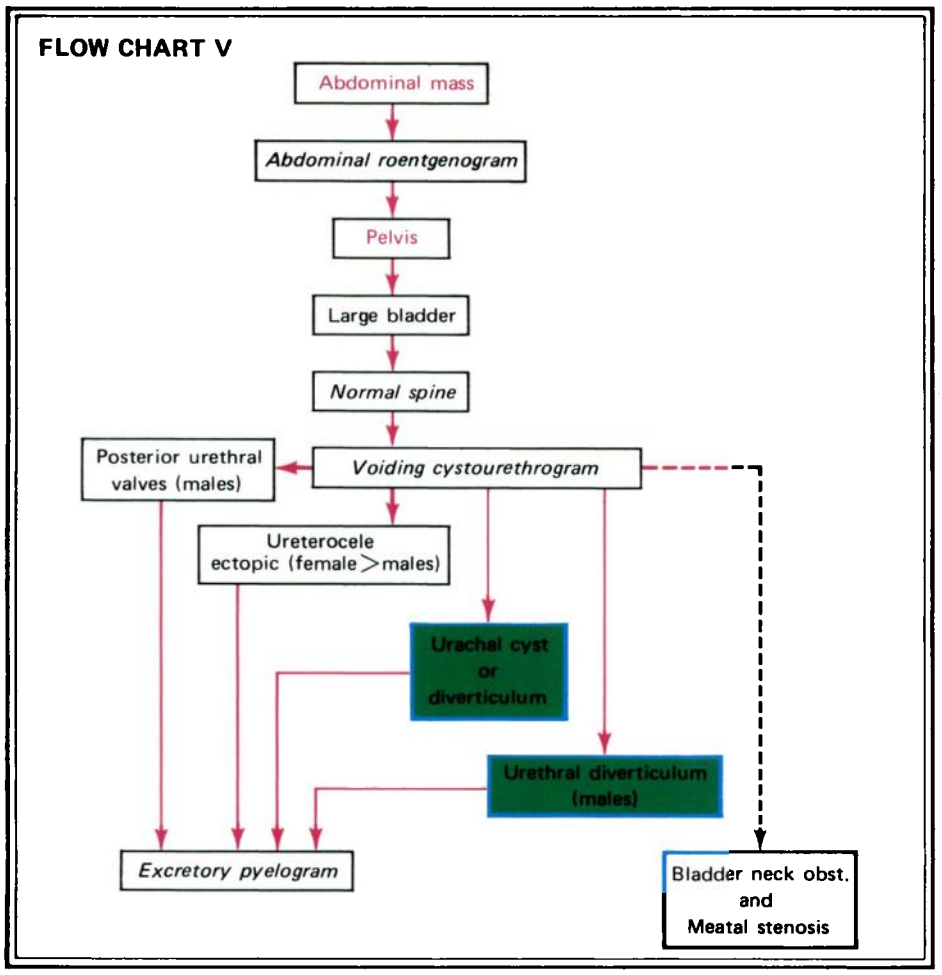
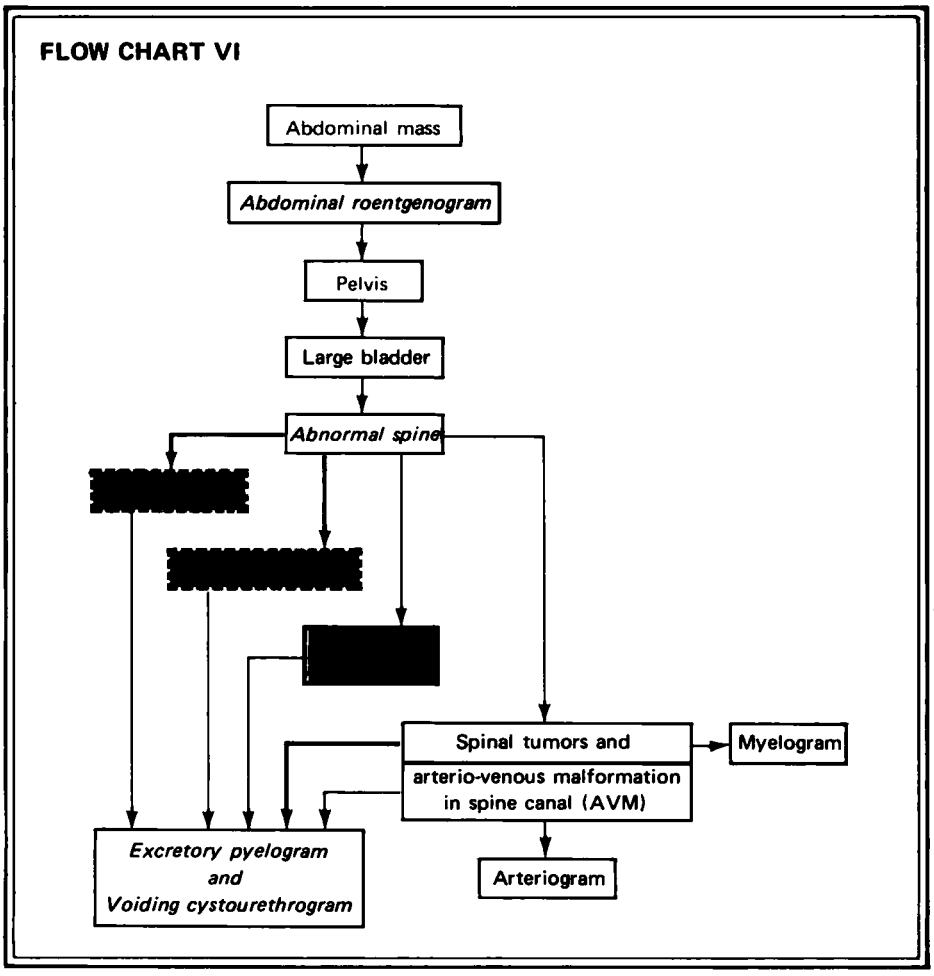


Fig. 8. (A.) The ring-like calcification (arrow) demonstrated at 12 days of life is typical of an adrenal hemorrhage as it contracts; the right kidney is displaced downward. (B.) Excretory urogram several months later shows the normal orientation of the right kidney and residual of calcification caused by the adrenal hemorrhage. (Courtesy of Dr. W. Berdon.)



tate and diffuse. (Fig. 9.) Thus, rather than immediately resorting to arteriography when the diagnosis is uncertain, it seems reasonable to observe a neonate with an adrenal mass for several days.[19] If curvilinear calcification and/or shrinkage appear, the mass can be identified as adrenal hemorrhage. In almost all patients, carefully staged clinical and radiologic evaluation leads to a correct diagnosis; arteriography is very rarely indicated.

The disease process of neuroblastoma has several hallmarks. The peak incidence is two years of age or younger, with approximately three-fourths of all tumors occurring during the first four years of life. In addition to the characteristic roentgenographic and pyelographic findings described above, evidence of metastases in the liver and/or skeleton is found in a large percentage of patients at the time of diagnosis. Radioactive isotope studies reveal skeletal metastases earlier than roentgenograms. Liver scans are adequate for evaluating liver metastases. Catecholamine determinations (such as vanillymandelic acid and homovanillic acid) in a 24-hour urine collection are increased in approximately 80-90 percent of children with neuroblastoma. Neuroblastoma cells in



the bone marrow are found in a high percentage of patients.

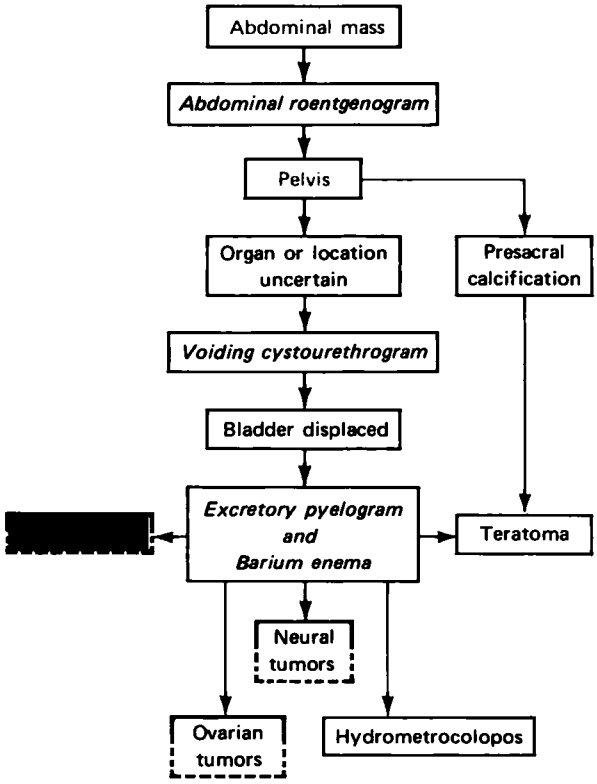
Survival is very poor in patients more than one year old, and it has not been improved by treatment with chemotherapy and/or radiation therapy.[20] In view of the extremely poor prognosis, angiography, lymphangiography, retroperitoneal air and other invasive procedures cannot be justified.

Adenoma, carcinoma and pheochromocytoma of the adrenal gland cause displacement of the kidney downward and occasionally laterally. Adrenal hormone studies, as well as clinical findings, provide information for accurate differentiation of these tumors.[21] Neovascularity on arteriography is not diagnostic; it has been found in benign as well as malignant tumors of the adrenal gland. Once a tumor has been detected by excretory pyelography, its nature may not be further characterized by arteriography. In childhood pheochromocytoma, with an incidence of multiple tumors as high as 30 percent, aortography is recommended for identification of additional tumors.[22]

OTHER RETROPERITONEAL MASSES

Other tumors such as lymphoma, tera-

FLOW CHART VII



toma, lymphangioma, rhabdomyosarcoma or lipoma may occasionally be found in the retroperitoneal space. Lymphomas are usually associated with peripheral adenopathy; a diagnosis is made from biopsies. Abdominal lymphangiography is still recommended by many investigators for staging Hodgkin's disease. Neural tumors may be associated with spine and/or rib changes. Teratomas can be identified by characteristic calcifications. Due to their fat content, lipomas appear as lucent masses on abdominal roentgenography, a lucency which is further exaggerated on bodygram and pyelogram.

PELVIC MASSES

An enlarged bladder is the most common cause of a pelvic mass, and may be found in patients with three types of clinical findings. Those with:

1. *Normal spines;*
2. *Abnormal spines;*
3. *Displacement of the bladder.*

Normal Spine

(FLOW CHART V)

In infants with normal spines, the pres-

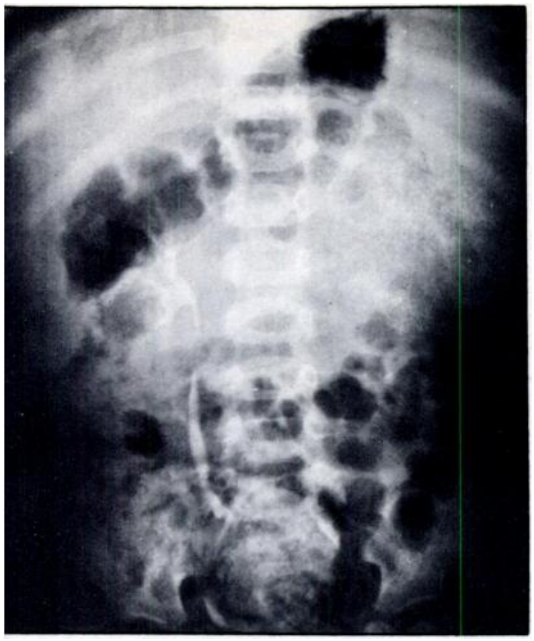


Fig. 9. A young child with neuroblastoma demonstrating diffuse punctate calcification in the left side of the abdomen adjacent to T-12—L-3. The left kidney is displaced downward. The right kidney and ureter are normal.

ence of a posterior urethral valve in males is the most frequent cause of an enlarged bladder.[23] When the abdominal roentgenogram shows a large bladder, the diagnostic study of choice is a voiding cystourethrogram, which defines a dilated prostatic urethra and a narrow stream distal to the external sphincter. (Fig. 10.) Lucent areas due to the valves are occasionally noted during the voiding study.

Rhabdomyosarcoma of the bladder, prostate[24] or vagina may sometimes initially present as enlargement of the bladder. Urgency and difficulty passing urine are characteristic. Hydronephrosis secondary to obstruction by tumor at the ureterovesicle junction may occasionally be the first sign of disease. The cystogram shows irregular defects caused by a tumor in the base of the bladder; a prostatic or vaginal tumor may also displace the bladder. (Fig. 11.) Roentgenographic evaluation includes excretory pyelography, cystourethrography, liver and skeletal isotope scanning, as

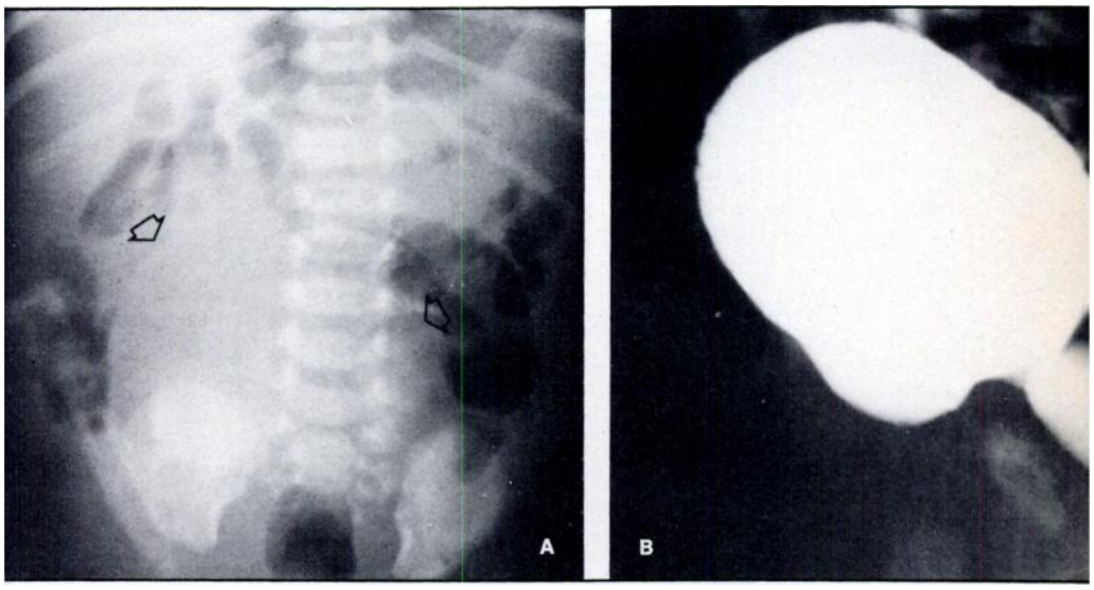


Fig. 10. A male infant in the first week of life had a poor voiding pattern; a large abdominal mass was found. (A.) Abdominal roentgenograms show that this mass originates in the pelvis and extends out of the true pelvis (arrows). (B.) A voiding cystourethrogram shows an enlarged bladder caused by a dilated prostatic urethra, due to posterior urethral valves, and a narrow stream distal to the external sphincter.

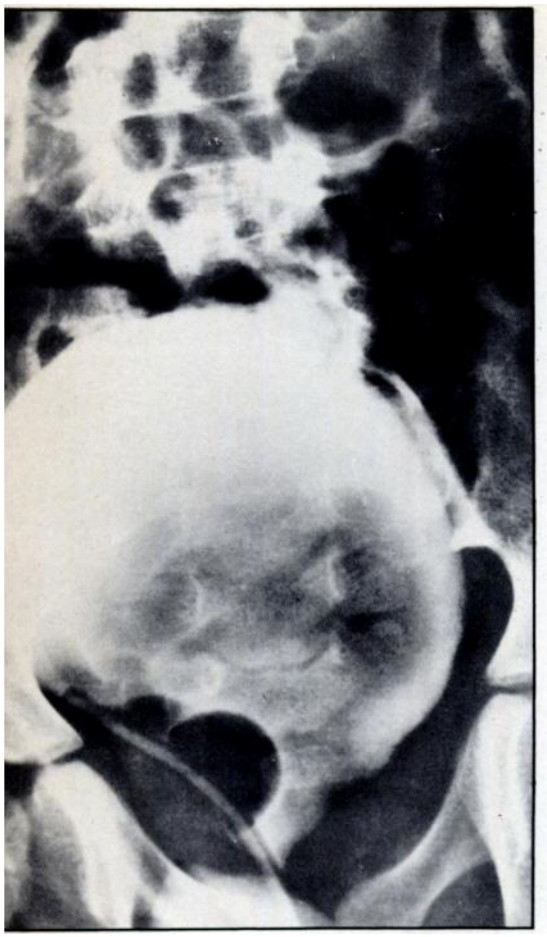


Fig. 11. A three-year-old boy with a large pelvic mass. The cystogram demonstrates a filling defect in the base of the bladder. The irregular pattern is characteristic of a rhabdomyosarcoma of the bladder or prostate.

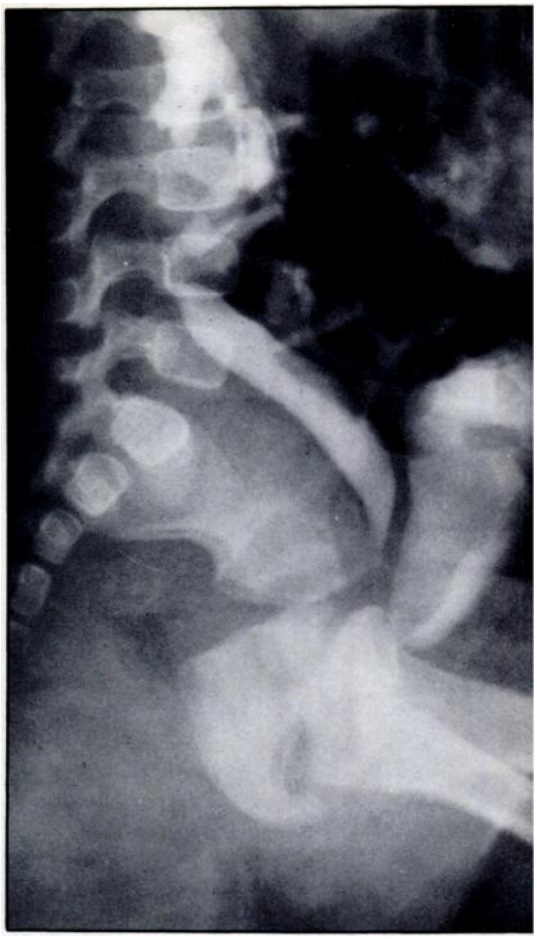


Fig. 12. A thirteen-month-old female with a soft tissue mass on her buttock since birth. Lateral view of the abdomen and buttock during an excretory pyelogram demonstates a large soft tissue mass extending beyond the coccyx. Calcification was identified on the roentgenogram but is not well demonstrated in this photograph. The bladder and ureter are displaced anteriorly with partial obstruction of the ureter causing moderate hydronephrosis. A barium enema showed anterior displacement of the rectosigmoid as well.

well as chest X-rays. Lymphangiography and arteriography do not contribute data that is essential to diagnosis or treatment.

Abnormal Spine

(FLOW CHART VI)

Abdominal roentgenography may reveal an isolated, large, neurogenic bladder in patients with abnormal spines. Spina bifidia indicates the presence of a meningocele, meningomyelocele or an anterior meningocele. The latter occasionally presents as a pelvic mass.

Spinal tumors and arteriovenous malformation also cause a large neurogenic bladder. Detection of a tumor requires close inspection of the spine to note flattening of the vertebral pedicles; scoliosis may also be a sign of a mass. Myelography is indicated to demonstrate the tumor. Excretory pyelography should be performed to evaluate the renal status.

Displacement of the Bladder

(FLOW CHART VII)

In the neonate, displacement of the bladder may be due to hydrometrocolpos, caused by imperforate hymen or atresia of the vagina. This diagnosis is readily made by physical examination.

Sacrococcygeal teratoma[25] may present as an external mass, although the major component is often within the pelvis, between the coccyx and the rectum. (Fig. 12.) On abdominal roentgenography, the rectum and bladder are displaced anteriorly; calcification within the teratoma is commonly seen. Further evaluation includes excretory pyelography to evaluate any possible secondary obstruction of the upper urinary tract and a barium enema to determine the amount of displacement of the lower colon and, thus, the extent of the mass. Teratomas found in children only a few months old have a low malignancy rate, compared to those detected in children more than a year old.

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