TOP 3 DIFFERENTIALS IN RADIOLOGY

Editor-in-Chief: William T. O’Brien, Sr., D.O.

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JAOCR About the Journal

Aims and Scope
The Journal of the American Osteopathic College of Radiology (JAOCR) is designed to provide practical up-to-date reviews of critical topics in radiology for practicing radiologists and radiology trainees. Each quarterly issue covers a particular radiology subspecialty and is composed of high-quality review articles and case reports that highlight differential diagnoses and important teaching points.

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TOP 3 DIFFERENTIALS IN RADIOLOGY
Editor: William T. O’Brien, Sr., D.O.

From the Editor
In this Issue ............................................................................................................................. 4
William T. O’Brien, Sr., D.O.

Differential-Based Case Reviews
Cystic and Solid Cortically Based Mass in an Adolescent with Seizures .................................................. 5
William T. O’Brien, Sr., D.O.

Band Appearance of Vertebral Bodies ....................................................................................... 8
Ganesh Joshi, MBBS, Christopher Cerniglia, D.O., M.Eng.

Gastroesophageal Junction Narrowing with Proximal Esophageal Dilation ........................................ 11
Rocky Saenz, D.O., Lauren Corley, D.O., Zack Franks, D.O.

Lower Limb Linear Bone Scintigraphy Activity ............................................................................ 14
Timothy McKnight, D.O.

JAOCR at the Viewbox
Congenital Absence of the Pericardium ..................................................................................... 18
Kyle Costenbader, MSIII, John P. Lichtenberger III, M.D.

Autosomal Recessive Polycystic Kidney Disease (ARPKD) .......................................................... 19
Sara A. Hunter, M.D., Brooke S. Lampl, D.O.

Median Arcuate Ligament Syndrome ....................................................................................... 20
Scott Sosin, D.O., Philip Orons, D.O.
Letter from the Editor

In this Issue

William T. O’Brien, Sr., D.O.
Editor-in-Chief, Journal of the American Osteopathic College of Radiology

“You can live to be a hundred if you give up all the things that make you want to live to be a hundred.”
-Woody Allen

“The past 8 years have been nothing short of incredible as we have watched the Journal grow from merely an idea back in 2011 to the exceptional educational and CME resource that it has become in 2019. I am forever indebted to the guest editors and contributors for volunteering their time and expertise to the college and the JAOCR, the editorial board for their insight in establishing the Journal content and format, and the staff at the AOCR office for their countless hours of behind-the-scenes work in coordinating each issue and its CME activities. I would also like to thank the staff at Anderson publishing for their editing support and production of each high-quality issue.

To say that the Journal will be in good hands with the incoming editor-in-chief would be a tremendous understatement. When it became evident that we needed to start looking for candidates to serve as editor, the Leadership Identification Committee came to the same recommendation that I had prepared—Dr. Daniel Wale, a nuclear medicine subspecialty-trained radiologist and faculty member at the University of Michigan. In addition to his active involvement in AOCR committees and educational programs, Dan’s acumen for publishing became readily apparent when he served as guest editor for a nuclear medicine subspecialty issue of the JAOCR. His enthusiasm, knowledge, experience, and editorial expertise make him a perfect fit as editor of the JAOCR for years to come.

In closing, I would like to thank the college for the many opportunities it has provided me over the years, especially the opportunity to serve as inaugural editor of the JAOCR. For the members of the AOCR, I encourage you to become involved in the college in any way feasible, whether it be through committee involvement, providing online or in person CME lectures, serving as guest editor for the JAOCR, or simply attending AOCR events. From my experience, the return on your investment will be ten-fold what you put into it.”
Case Review: O’Brien

Cystic and Solid Cortically Based Mass in an Adolescent with Seizures: A Case-Based Illustrative Review

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Case Presentation
A 14-year-old girl presented with intermittent episodes of dizziness, staring spells, and “odd smiling episodes” that had been increasing in frequency over the past 4 years. She denied headaches, visual, motor, or sensory symptoms associated with the episodes. There was no overt “post-ictal” state. The patient was referred for a brain MRI for suspected seizures (Figure 1).

FIGURE 1. Axial (A) and coronal (B) T2/FLAIR images show a cortically based right temporal lobe mass with hyperintense solid, small hypointense cystic components, and mild adjacent parenchymal edema. T1 pre- (C) and postcontrast (D) images show enhancement of the solid components, nodular along its superficial margin (arrow, D). Mild gyral expansion is seen on all images.
Case Review: O’Brien

Key Imaging Finding(s)
- Cystic and solid cortically based mass in an adolescent

Differential Diagnoses
- Ganglioglioma
- Pleomorphic xanthoastrocytoma
- Dysembryoplastic neuroepithelial tumor

Discussion
Seizures are a common clinical indication for advanced neuroimaging in pediatric patients. The goal of imaging is to identify a potential epileptogenic focus, especially in cases that are refractory to medical management. Common etiologies of epileptogenic foci include congenital, acquired, and neoplastic lesions. Although any neoplasm may result in seizures, neoplasms particularly prone to epilepsy in children, adolescents, and young adults include ganglioglioma, pleomorphic xanthoastrocytoma (PXA), and dysembryoplastic neuroepithelial tumor (DNET).

Ganglioglioma
Ganglioglioma is a mixed neuroepithelial tumor with neoplastic ganglion and glial components. It is cortically based, most often occurs in the temporal lobes, and represents the most common neoplastic cause of refractory epilepsy in children and young adults. Its peak presentation occurs in the second decade of life.

The imaging appearance often mimics its histology. The vast majority of lesions are low-grade (WHO grade 1) and present as solid or cystic and solid cortically based masses. On MRI, the solid component is iso- to hypointense compared to gray matter on T1 and hyperintense on T2/FLAIR imaging. Both enhancement and calcification are relatively common, occurring in about 50% of cases. Enhancement of solid lesions may be heterogeneous or atypical imaging appearance with ill-defined signal abnormality or areas of necrosis. Surgical intervention is the treatment of choice and may be curative with gross total resection. Radiation therapy or radiosurgery is typically reserved for recurrent, aggressive, or unresectable cases.

Pleomorphic Xanthoastrocytoma
Similar to ganglioglioma (although less common), PXA is a cortically based tumor that most commonly occurs in the temporal lobes and may be a cause of refractory partial complex seizures in adolescents and young adults. It is derived from astrocytes and is considered a WHO grade II tumor, although anaplastic features may be present in rarer higher-grade lesions.

The characteristic imaging appearance of PXA is that of a cystic mass with an avidly enhancing mural nodule. The nodule typically abuts the overlying meninges, which are often thickened and may show dural enhancement, which is a useful discriminator when present. The solid component has variable density on CT and is iso- to hypointense to gray matter on T1 and variably hyperintense on T2/FLAIR imaging. Calcification may be seen, whereas hemorrhage is rare. A less...
common imaging appearance is that of a mainly solid tumor with varying degrees of cystic change. The degree of surrounding edema may vary from none to marked. Its peripheral location and slow-growing nature may cause bony remodeling.5,6

Surgical resection is the preferred treatment with gross total resection often being curative. Residual or recurrent tumor may be treated with additional resection and/or radiation therapy.

Dysembryoplastic Neuroepithelial Tumor

DNET is a low-grade (WHO grade I) mixed tumor with glial and neuronal components. It is a cortically based neoplasm that most commonly occurs in the temporal lobes. In combination with cortical dysplasia that is often present in association with the lesion, patients commonly present in the first and second decades of life with medically refractory epilepsy.

The characteristic imaging appearance is that of a circumscribed, wedge-shaped, “bubbly” or multicystic-septated cortically based mass (Figure 5). The noncystic component is often iso- to hypointense compared to the cortex on T1 and iso- to hyperintense on T2/FLAIR imaging. There is typically little mass effect and no significant surrounding vasogenic edema. Enhancement may occur in about one-third of cases with calcifications being less common. The associated cortical dysplasia often demonstrates blurring of the gray-white junction with more ill-defined and tapered margins. Given the slow-growing nature of the benign tumor, overlying bony remodeling may be seen with peripheral lesions.7

Gross total surgical resection of the lesion and associated cortical dysplasia is often curative.8

Diagnosis

Pleomorphic xanthoastrocytoma

Summary

Advanced neuroimaging is commonly performed in children and adolescents to identify a potential epileptogenic focus. The most common neoplasms prone to medically refractory epilepsy in the pediatric population include ganglioglioma, PXA, and DNET. Although the imaging characteristics of these lesions overlap, secondary or specific signs may often favor one lesion over another. Therefore, it is important for radiologists to have a basic understanding of the differentiating features associated with these epileptogenic tumors to help guide appropriate management.

REFERENCES

Band Appearance of Vertebral Bodies: A Case-Based Illustrative Review

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Case Presentation
A 43-year-old man presented with chronic progressive back pain. Thoracic spine radiographs were performed (Figure 1).

FIGURE 1. Lateral thoracic spine radiograph shows sharply demarcated sclerosis abutting the vertebral body endplates of the vertebrae with central relative lucency, resulting in the classic “sandwich vertebra” appearance. Vertebral body heights and morphology are normal.
Key Imaging Finding(s)

- Band appearance of vertebral bodies on radiographs

Differential Diagnoses

- Osteopetrosis
- Renal osteodystrophy
- Osteoporosis

Discussion

Sclerotic and lucent bands in multiple thoracic and lumbar vertebral bodies result in characteristic imaging findings on radiographs. These findings can be observed in patients with osteopetrosis, renal osteodystrophy, and osteopenia. The purpose of this article is to review the imaging features of these entities and how to differentiate one from another based on radiographic findings.

Osteopetrosis

Osteopetrosis is a hereditary skeletal dysplasia that results in extremely dense bones throughout the skeleton. This disorder is characterized by decreased bone resorption with decreased osteoclastic activity, resulting in increased bone mass. There are two forms of the disorder – the infantile (malignant) form, which is transmitted in an autosomal recessive pattern, and the adult (benign) form, which is transmitted in an autosomal dominant pattern. The infantile form typically leads to stillbirth or early death, while the adult form, also known as Albers-Schonberg disease, is less severe with a delayed manifestation. The adult form is most often discovered incidentally on radiographs in otherwise asymptomatic young or middle-aged adults. Less often, patients present with scoliosis, arthritis, fractures, or osteomyelitis related to the underlying disorder.

On imaging, osteopetrosis manifests as increased density within the medullary portion of the bone with relative sparing of the cortices. Autosomal dominant osteopetrosis (ADO) has two phenotypic variants – uniform sclerosis of the long bones, spine, and pelvis in the type I ADO variant, and the classic “sandwich vertebra” appearance with sharply marginated, densely sclerotic endplates (Figure 1) in the type 2 ADO variant. The “bone within bone” appearance involving the skull base and pelvis is also characteristic of type 2 ADO (Figure 2).  

Renal Osteodystrophy

Renal failure may present with a variety of musculoskeletal complications, either related to the disease itself or its treatment. Renal osteodystrophy results from altered metabolism associated with...
renal failure, manifesting as a combination of osteomalacia and secondary hyperparathyroidism.\textsuperscript{4} Osteomalacia refers to insufficient or delayed mineralization of osteoid tissue in adults, resulting in osteopenia. Twenty percent of patients with renal osteodystrophy, however, present with osteosclerosis involving ribs, pelvis, and spine.\textsuperscript{5} The radiographic picture of renal osteodystrophy can be confusing, since one often has overlapping findings of osteomalacia and hyperparathyroidism. In children, one may also see the rachitic findings overlapping with these two processes.

Radiographically, renal osteodystrophy commonly presents as sclerotic bands along superior and inferior endplates of vertebral bodies with intervening central lucency. Alternate sclerosis and lucency with ill-defined margins give rise to the characteristic “rugger jersey spine” appearance (Figure 3).

**Osteoporosis**

Osteoporosis is the most common metabolic disorder in elderly patients. It is characterized by decreased bone mass with microarchitectural deterioration of bone tissue, resulting in increased fragility. Osteoporosis affects millions of people worldwide with an estimated 2.3 million fractures in the United States and Europe alone.\textsuperscript{6} There are two types of osteoporosis – primary and secondary. Primary osteoporosis usually occurs in elderly patients with cumulative bone loss from aging and hormonal changes, while secondary osteoporosis results from various medical conditions and certain medications.

On radiographs, the main imaging features of osteoporosis include increased radiolucency and cortical thinning. Osteopenia is a frequent term used to describe the radiolucency on radiographs, which results from resorption and thinning of the bony trabeculae.\textsuperscript{7} The radiolucency commonly is most evident along the superior and inferior endplates of the vertebral bodies, resulting in the “reverse rugger jersey” appearance of the spine (Figure 4).

**Diagnosis**

**Osteopetrosis**

**Summary**

Radiography is the most commonly performed imaging modality in the initial evaluation of back pain. When a band appearance of the vertebral bodies is noted, osteopetrosis, renal osteodystrophy, and osteoporosis are the most likely etiologies. Although differences may be subtle, there are a few specific features that may favor one entity over another. Radiologists should be aware of these characteristic imaging findings to suggest the most likely diagnosis and help guide appropriate management.

**REFERENCES**

Gastroesophageal Junction Narrowing with Proximal Esophageal Dilation: A Case-Based Illustrative Review

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Case Presentation
A 58-year-old woman presented with long-standing dysphagia without weight loss or hematemesis. She was subsequently referred for an upper GI (UGI) examination (Figure 1).

FIGURE 1. An oblique frontal view from an UGI examination reveals a dilated esophagus that tapers to a “bird’s beak” appearance distally at the gastroesophageal junction. Reprinted with permission from Top 3 Differentials in Radiology, 2nd ed. (p. 78-79) by O’Brien WT, 2018, New York, NY: Thieme.
Key Findings
- Gastroesophageal junction narrowing with esophageal dilation

Differential Diagnosis
- Esophagitis
- Achalasia
- Esophageal carcinoma

Discussion
Dysphagia is a common indication for an UGI fluoroscopy examination or esophagography. The goal of imaging is to identify mucosal irregularity, ulceration, or stricture. There are numerous causes of distal esophageal strictures with the most common etiologies being esophagitis from a variety of causes, achalasia, and malignancy. Classically, malignant strictures have a recent onset of rapidly progressive dysphagia and weight loss, while benign strictures present with long-standing, nonprogressive dysphagia. The most common etiology overall of distal esophageal narrowing results from gastroesophageal reflux disease (GERD).

Esophagitis
Inflammation of the esophagus is most commonly caused by GERD but can also be secondary to infection, chemotherapy, caustic ingestion, or radiation. Reflux esophagitis or GERD affects an estimated 40% of adults. Common symptoms include esophageal dysphagia, substernal or epigastric pain, food regurgitation, and dry cough. Patients with hiatal hernias are at an increased risk of having reflux. During fluoroscopy, contrast is seen refluxing into the esophagus from the stomach. Chronic reflux can result in luminal narrowing, which appears smooth and concentric, typically 1 to 4 cm in length (Figure 2). A more severe complication of GERD is intestinal metaplasia of the lower esophageal mucosa, called Barrett esophagus, which rarely can progress to adenocarcinoma. Treatment for GERD includes lifestyle modifications, pharmaceutical treatment with proton pump inhibitors, H-2 receptor blockers, or antacids. In patients with a hiatal hernia, fundoplication may also alleviate symptoms.

Achalasia
Achalasia is a motility disorder of the esophagus with absent primary peristalsis resulting in impaired relaxation of the lower esophageal sphincter (LES). Patients present with symptoms of dysphagia to solids and liquids, chest pain, food regurgitation, and are typically middle-aged. Achalasia is categorized as primary (idiopathic) or secondary, which results from destruction of the plexus by an infiltrating tumor (commonly known as pseudoachalasia) or infections such as Chagas disease. Idiopathic achalasia occurs from destruction of the myenteric plexus neurons in the LES, resulting in failure to relax. Esophagography classically shows a dilated esophagus and symmetrical tapering near the LES with a characteristic “bird-beak” appearance (Figures 1 and 3). Manometric testing is the gold standard for diagnosing primary achalasia. Idiopathic achalasia is more definitively treated surgically, with graded pneumatic balloon dilation or with Heller myotomy, but can be conservatively managed with calcium channel blockers or Botulinum toxin.

Esophageal Carcinoma
Esophageal carcinoma is most often squamous cell carcinoma (SCC), followed by adenocarcinoma. SCC is typically seen in the proximal two-thirds of the esophagus, while adenocarcinoma...
is more common in the distal one-third of the esophagus. Risk factors for esophageal SCC include tobacco use, alcohol consumption, achalasia, and long-standing esophagitis. Adenocarcinoma is more closely related to GERD and Barrett metaplasia. Patients with pseudo-achalasia from tumor infiltration present with an abrupt onset of symptoms, compared with benign etiologies. On fluoroscopy studies, carcinomas usually appear as eccentric or asymmetric wall thickening with irregular nodular mucosa, abrupt “shouldering,” and mass effect (Figure 4). The diagnosis is confirmed with endoscopy and biopsy.

Diagnosis

Achalasia

Summary

Upper GI and esophagography examinations are commonly performed in the setting of dysphagia with the primary goal of identifying potentially treatable areas of mucosal irregularity, ulceration, or stricture. Distal esophageal strictures are relatively common and are most often due to esophagitis, achalasia, or esophageal carcinoma. Each entity has fairly characteristic imaging findings, which often can be readily identified on fluoroscopic studies. Therefore, it is imperative that radiologists recognize these findings, especially in terms of distinguishing benign from malignant strictures, to help guide appropriate management and follow-up.

References

Lower Limb Linear Bone Scintigraphy Activity: A Case-Based Illustrative Review

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Case Presentation

An 18-year-old lacrosse player presented with persistent bilateral lower extremity pain, for which nuclear medicine scintigraphy was performed (Figure 1).

FIGURE 1. Tc-99m hydroxydiphosphonate (HDP) delayed anterior-posterior (AP, upper) and lateral (lower) planar images of the legs demonstrate linear patchy uptake involving the bilateral tibial posterior cortices. Perfusion and blood pool phase images (not shown) were normal.
Key Imaging Finding
Lower limb linear long bone activity

Differential Diagnoses
Medial tibial stress syndrome (shin splints)
Fracture
Osteomyelitis

Discussion
Lower extremity pain is a common complaint across age groups. In many situations, the clinical picture may be suggestive, but is often not definitive for a specific diagnosis. Radiographs are often the first study of choice but may be normal or equivocal. Bone scintigraphy has long been a popular and accessible next imaging option of choice and may provide useful information as to the etiology of lower extremity pain, impacting patient treatment and management options. Common conditions demonstrating a linear pattern of lower limb long bone activity include medial tibial stress syndrome (MTS), fractures, and osteomyelitis, detailed below. Other uncommon etiologies, such as venous stasis and hypertrophic pulmonary osteoarthropathy, are not discussed in this case review.

Medial Tibial Stress Syndrome
Medial tibial stress syndrome (MTS), colloquially known as shin splints, results from repetitive stress injury to the leg. The clinical presentation is exertional leg pain localized along the posterior medial border of the middle and/or distal thirds of the tibias. Radiographs are typically normal (Figure 2). Tc-99m bone scintigraphy has long been a useful tool in confirming this syndrome and excluding more severe pathologic mimics. Abnormal delayed phase uptake is typically seen in most, but not all, proven cases of MTS.\(^1\) Perfusion and blood pool phases are typically normal.

Three patterns of uptake are closely associated with MTS: superficial uptake along the anterior and posterior tibial cortices (double stripe sign),\(^2\) localized middle to distal tibial posteromedial...
cortex activity (Figure 1), and tubular pattern of tibial activity. MTS most commonly involves at least one-third of the tibial shaft, is usually bilateral, and is often asymmetric and heterogenous.

Histological changes of MTS include osteocyte loss, enlarged lacunae, disrupted lamellar structure, and periosteal thinning or thickening. Interestingly, the extent and severity of histologic periosteal changes do not correlate with the intensity of uptake. This disparity has been suggested to represent differences between early and late/reparative phases of the disease process with abnormal activity preceding significant histologic changes.

MRI can provide a useful adjunct assessment in equivocal cases with a progressive spectrum of findings, including normal or linear increased fluid signal periosteal edema with or without increased fluid signal marrow (Figure 3). The presence of intracortical abnormal signal crosses into the imaging patterns of occult stress fracture. In this clinical setting, the extent and severity of MRI findings has been shown to correlate with the likelihood of response to conservative treatment and time to return to sports activity.

Fracture
Traumatic and stress fractures present with a fusiform or linear appearance (Figure 4). Stress fractures more commonly are transversely oriented to the cortex but longitudinal or oblique orientations may occur in about 10%. Combination of hyperperfusion, hyperemia, and delayed activity is typical, and in a young healthy patient appears within 24 hours of the inciting event. Delayed images at 72 hours may provide increased sensitivity, especially for elderly patients who may have delayed reparative response. Overall sensitivity of radionuclide imaging approaches 100%. The pattern and severity of the fracture will determine the need for surgical vs. non-surgical reduction and immobilization to facilitate healing. Uncomplicated fracture activity will persist for at least 6 months with most resolved by 2 years, excluding chronic nonaligned injuries, which may have indefinitely prolonged activity.

Osteomyelitis
Infection shares the hyperperfusion, hyperemic, and intense delayed phase activity that is seen in fractures and can be focal, fusiform, linear, or diffuse in distribution (Figure 5). The scintigraphic appearance has a high accuracy (> 90%) in the setting of high clinical index of suspicion, but a low specificity if the clinical presentation is equivocal. Additional studies, such as radiographs, MRI, indium-111-labeled autologous-leukocytes, or biopsy may be required for diagnostic confirmation. The high negative predictive value of a normal bone scintigraphy represents its best utility. Treatment
may include antibiotics and/or surgical debridement. Untreated or chronic osteomyelitis may result in progressive bone destruction, abscess formation, and sepsis with significant increase in mortality.

**Diagnosis**

Medial tibial stress syndrome (shin splints)

**Summary**

Bone scintigraphy is a useful and commonly utilized tool for assessing lower extremity pain. While the patterns of linear uptake among the differential diagnoses may overlap to some degree, subtle differences may be apparent, which when combined with the clinical picture, provide clues to the underlying diagnosis. Correlation with additional imaging – often radiographs – is helpful in suggesting the most likely diagnosis. Often, the best practical application of bone scintigraphy is when it is normal, providing a high degree of confidence in excluding each of these processes. The radiologist should have a general understanding of bone scintigraphy’s role in the workup of a patient with lower extremity pain, be familiar with the common pathologic entities and their presenting patterns on scintigraphy, and be cognizant of the impact of imaging findings in terms of patient management and outcomes.

**References**


Congenital Absence of the Pericardium

A 24-year-old asymptomatic man presented for preoperative chest radiography. Posteroanterior chest radiograph (A) shows levoposition of the heart with loss of the right heart border, superolateral displacement of the cardiac apex, and lucency between the heart and diaphragm. Axial contrast-enhanced CT (B) shows no identifiable pericardium or pericardial fat, consistent with congenital absence of the pericardium (CAP).

CAP is a rare diagnosis in which the pericardium fails to form, most commonly due to premature atrophy of its vascular supply. There is a male predominance of 3:1. CAP may be complete or partial with the most common presentation being a left-sided partial absence. Complete CAP is usually clinically insignificant. However, partial absence of the pericardium may present with chest pain or arrhythmia, or be complicated by fatal herniation and incarceration of the ventricles or atrial appendages.1

Chest radiography findings of CAP include levoposition of the heart, a posteriorly and superiorly displaced cardiac apex, and abnormally interposed lung parenchyma. Definitive diagnosis of CAP is made on cardiac CT or MRI and demonstrates three findings: 1) failure to visualize the hypointense pericardial line between myocardium and pericardial fat; 2) levoposition of the heart; and 3) abnormal interposition of lung parenchyma between the heart and diaphragm or in the aorto-pulmonary space. Additionally, transesophageal echocardiogram may disclose paradoxical motion of the interventricular septum, falsely enlarged cardiac chambers due to the shifting heart, and absent systolic separation of the pericardial layers.2 Treatment is surgical and generally reserved for patients who have debilitating symptoms or herniation.

REFERENCES
Autosomal Recessive Polycystic Kidney Disease (ARPKD)

An 8-year-old girl with autosomal recessive polycystic kidney disease (ARPKD) presented for routine follow-up. Renal ultrasound demonstrated enlarged, echogenic kidneys with lack of corticomedullary differentiation and innumerable small cysts (A and B). Corresponding CT examination demonstrated lobulated enlargement of the kidneys with poorly enhancing hypodense parenchyma (C). The liver and biliary system were normal.

ARPKD is an inherited cystic renal disease, occurring in approximately 1:20,000 births, relating to a mutation of the polycystic kidney and hepatic disease 1 (PKHD1) gene on chromosome 6p. Pathologic findings are characterized by nonobstructive, symmetric renal collecting system ectasia with interstitial fibrosis. Patients most commonly present in the perinatal period with respiratory distress from pulmonary hypoplasia and/or renal insufficiency.1,2 Our patient initially presented at 4 years of age with multiple urinary tract infections and abdominal masses palpated on physical exam (corresponding to enlarged kidneys).

Patients with ARPKD also develop congenital hepatic fibrosis, characterized by hepatic biliary duct ectasia and fibrosis of the portal tracts. Although an inverse relationship between the severity of the renal and hepatic disease has been considered, a more recent study suggests that they may be independent factors. In general, children presenting early in life with severe renal disease often have relatively mild hepatic disease and those who present late with renal disease often have more severe hepatic disease.1,2 Treatment is primarily supportive with control of portal hypertension. Some patients require hepatic and/or renal transplantation. Overall prognosis is variable, depending on the severity of the renal disease, with the worst prognosis in the perinatal form.

REFERENCES
Median Arcuate Ligament Syndrome

A 32-year-old woman presented with upper abdominal pain and recent weight loss. She underwent CT angiography and conventional angiography demonstrating focal extrinsic compression and associated downward deflection of the celiac trunk (A, arrow).

Median arcuate ligament syndrome (also known as celiac artery compression syndrome, celiac axis syndrome, celiac trunk compression syndrome, or Dunbar syndrome) is a rare disorder characterized by chronic, recurrent abdominal pain related to compression of the celiac artery by the median arcuate ligament (a muscular fibrous band of the diaphragm). It usually presents in patients 20 to 40 years of age with symptoms of abdominal pain, weight loss, and abdominal bruit, and affects women far more frequently than men (35:1). It is unclear whether symptoms occur from vascular compromise or from compression of the celiac plexus. CT angiography and conventional angiography are considered the gold standard imaging modalities.

Imaging findings are characterized by a hooked appearance of the celiac axis due to superior extrinsic compression, frequently with associated poststenotic dilation (B, arrow). Functional stenosis may lead to collateral formation between the superior mesenteric and celiac arteries, most notably through hypertrophy of the pancreaticoduodenal arcades (C, arrow). When indicated by severity of symptoms, therapy is surgical decompression or bypass.

The characteristic appearance of the stenosis, absence of associated disease of the superior mesenteric artery, as well as the younger age of the patient, distinguishes this syndrome from the primary differential of atherosclerotic disease. It is noted that extrinsic compression of the celiac artery may also be seen in up to 50% of asymptomatic patients if imaging is acquired during the expiratory phase. Therefore, imaging for accurate diagnosis should ideally be performed in the end-inspiratory phase.

References