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**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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Letter from the Guest Editor

In this Issue

Mark Guelfguat, D.O.

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During the last few decades, cardiac imaging has evolved from relying on radiographs and fluoroscopy to a discipline primarily based on advanced cross-sectional imaging and molecular imaging techniques. Capitalizing on innovations, technology and unique techniques, this field unites previously diverse medical specialties and areas of expertise. It is the pleasure of the authors to provide the readers with glimpses of the spectacular spectrum of this distinguished subspecialty. The purpose of this issue is to deliver an approach to identifying and understanding common and not-so-common cardiac entities found in radiological practice.

Drs. Poletto, Mallon, Stevens, and Avitabile provide an impressive review of aortic arch and pulmonary artery variants. This work supplies state-of-the-art imaging illustrations and serves as a comprehensive reference guide to anyone involved in imaging congenital thoracic vascular diseases.

The current state of coronary CT angiography for management of acute chest pain in the emergency department is skillfully summarized by Drs. Rydzinski and Weg. This deft review not only provides the up-to-date clinical rationale for performing these examinations, but also equips the reader with its intricate technical background and imaging interpretation criteria.

A challenge of abnormal myocardial enhancement as commonly encountered in daily practice is discussed by Drs. Legasto and Waite. This impressive case review defines a framework of the first line MRI differential diagnoses pertinent to this phenomenon.

Drs. Newman and Meisner have masterfully reviewed the importance of a calcified mitral annular mass. Surprising nuances of imaging and clinical characteristics await the reader in this case review.

An interesting Viewbox article by Dr. Kim provides an insightful snapshot of hypertrophic cardiomyopathy. An intriguing article in this rubric by Dr. Salamon features surprising facets of myocardial noncompaction.

On behalf of the authors, I would like to express sincere gratitude to the AOCR Board, Journal, reviewers, editors, and publishers for their tireless work and support of the radiological community. Our special thanks go to Dr. William O’Brien for his guidance and encouragement. This issue would have been impossible without his energizing motivation. As a result of Dr. O’Brien’s vision and unwavering perseverance, the standards of the JAOCR continue to soar each year.

As I write this preface, I reflect on long hours spent by the authors working on this issue. I hope this work will provide the reader with greater insight into the complexities and benefits of cardiac imaging.
Aortic Vascular Rings and Pulmonary Sling, Poletto

Erica Poletto, M.D., Mary G. Mallon, M.D., Randy M. Stevens, M.D., Catherine M. Avitabile, M.D.

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Anatomic variants of the aortic arch and its branches are relatively common, with an estimated prevalence of 0.5% to 3%. The majority of these variations are of no clinical significance and are detected incidentally. However, several of these anatomic variants, grouped together under the term vascular ring, can produce respiratory symptoms or dysphagia due to encirclement and extrinsic compression of the trachea or esophagus, respectively. The contributors to the ring can include the aortic arch, arch vessels, or the ligamentum arteriosum, which is the fibrous remnant of the ductus. The Table presents a list of aortic variants that may be considered vascular rings.

Anatomic variation of the pulmonary artery and its branches is rarer than that of the aorta. Pulmonary arteries may be hypoplastic or absent, in isolation or associated with congenital heart disease. A pulmonary sling is a specific variant in which the left pulmonary artery arises from the right pulmonary artery instead of the main pulmonary artery. This abnormality can present with respiratory symptoms, similar to a vascular ring.

This review will discuss many common and rarer anatomic variants of the aortic arch and pulmonary arteries, with an emphasis on imaging modalities for vascular evaluation and their role in clinical decision-making and surgical repair.

Embryology

Any discussion of vascular rings and pulmonary artery slings must begin with a review of the embryology of the aortic arches, as this will help the radiologist or clinician understand potential anatomic abnormalities. The aortic arch system develops from the appearance and persistence or disappearance of six paired vessels originating from the embryonic foregut that connect the truncal aortic sac (the most distal part of the embryonic heart tube giving rise to the ascending aorta and the branch pulmonary arteries) to the paired dorsal aortae. The left 3rd arch gives rise to the left common carotid artery, while the left 4th arch develops into the definitive aortic arch between the left common carotid and the left subclavian artery. Ducti arteriosi arise from paired 6th arches. Therefore, in the typical left aortic arch, dissolution of the right 6th aortic arch (ductus arteriosus) and the right dorsal aorta proximal to the origin of the right 7th intersegmental artery (precursor to the right subclavian artery), results in the typical branching pattern.

While the 1st and 2nd arches become arteries of the face and ears, and the 5th arch usually disappears, the paired 3rd, 4th, and 6th arches as well as the paired 7th intersegmental arteries contribute to this “totipotent arch.” In the typical left aortic arch with normal branching pattern, the innominate artery originates from the right branch of the truncus arteriosus, with the right 3rd arch giving rise to the right common carotid artery. The right subclavian artery is derived from contributions from the right 4th arch, the right 7th intersegmental artery, and the right dorsal aorta. The left 3rd arch gives rise to the left common carotid artery, while the left 4th arch develops into the definitive aortic arch between the left common carotid and the left subclavian artery. Ducti arteriosi arise from paired 6th arches.

When certain arches fail to regress, they can persist into the post-natal period, giving rise to a multitude of variants. When the left and right 4th arches
persist to connect the truncoaortic sac to the dorsal aorta, the result is a double aortic arch. Both arches can persist in their entirety, or a portion of one of the arches (typically the left arch) can become atretic.

When the left 4th arch regresses in its entirety, a right aortic arch results. With a right aortic arch, the left subclavian artery may arise from a left innominate artery (called mirror-image branching, because it is the vertical mirror-image of standard anatomy). Alternatively, the left subclavian artery may arise as a separate branch from the distal aspect of the right-sided aortic arch; this aberrant subclavian artery must cross the midline posterior to the esophagus to achieve its leftward course.

When the dorsal aorta contralateral to the aortic arch persists, and the ipsilateral dorsal aorta regresses, the result is a circumflex aorta. Right circumflex aortic arch with left descending aorta is more common than left circumflex aortic arch with right descending aorta. The subclavian artery contralateral to the side of the aortic arch may arise from the innominate artery or aberrantly.

If the aortic arch arises from the left or right 3rd arch, instead of the 4th arch, a high-lying aortic arch results. While this

### Table. Aortic arch variants

<table>
<thead>
<tr>
<th>Vascular Ring</th>
<th>Not a Vascular Ring</th>
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<tbody>
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<td>Double aortic arch</td>
<td>Left aortic arch with normal branch pattern</td>
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<td>Patent left and right arches</td>
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<td>Right aortic arch with mirror image branching and retroesophageal left ligamentum arteriosum (rare)</td>
<td>Direct origin of vertebral artery from arch</td>
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<td>Left aortic arch with retroesophageal diverticulum of Kommerell and aberrant right subclavian artery (rare)</td>
<td>Cervical arch (in isolation only; can be vascular ring if found in conjunction with variant from left column)</td>
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</tbody>
</table>

**FIGURE 1.** Schematic of 6 paired arches and 7th intersegmental artery.
is called a *cervical arch*, the arch is typically at or just above the clavicular heads.

While the embryology of pulmonary artery sling is unclear, it is likely due to a failure of the proximal left 6th arch (precursor of the proximal pulmonary arteries) to join the distal lung buds. A secondary vessel originates from the right side to join the distal lung buds.

**Clinical Presentation**

Clinical presentation of patients with vascular rings is variable and is related to the degree of tracheal compression. Infants often present with stridor, with significant increase in respiratory distress in infancy. Respiratory symptoms or dysphagia. Patients with pulmonary artery slings are often symptomatic with stridor and respiratory distress in infancy. Respiratory symptoms may also be attributed to concurrent hypoplasia or dysplasia of the trachea, with up to 65% of patients having complete tracheal cartilaginous rings.1

**Imaging of Vascular Rings and Slings**

Because patients with vascular rings and slings often present with respiratory symptoms, the imaging evaluation often starts with a two-view chest radiograph. In up to 95% of patients with a vascular ring/sling, an abnormality is visible on chest radiographs.4 However, many factors may limit accurate assessment of the trachea on the frontal view, particularly in infants: An atretic or hypoplastic arch may not be large enough to cause mass effect; the large thymus may obscure the aortic arches; and radiographs may be obtained during expiration and/or with rotation that limits evaluation of the position of the trachea.

Patients with vascular rings can also present with dysphagia, prompting a fluoroscopic upper gastrointestinal (UGI) study. This study can contribute important information to the characterization of a vascular ring/sling. Indentation on the posterior esophagus on a lateral view suggests a vascular ring, but does not distinguish the type. While imaging in the frontal projection can identify the sided-ness of the aortic arch(es), the indentation on the esophagus from the aortic arch(es) can be subtle. Additional imaging is often required to completely characterize the nature of the aortic arch. Indentation on the anterior esophagus on a lateral view suggests the presence of pulmonary sling.

Once suspicion of vascular ring/sling is raised, evaluation typically progresses to cross-sectional imaging. Echocardiography is a noninvasive imaging modality that imposes no risks of radiation exposure or contrast administration. Echocardiography provides information about cardiac function and direct visualization of cardiac and vascular anatomy. The sensitivity of echocardiography for evaluating vascular rings/slings varies widely (30% to 100%), depending on the sonographic window available and sonographer skill.4 Echocardiography has the potential to determine arch sidedness and delineate the branch pattern (Figure 2), but is limited regarding tracheal or esophageal compression by the surrounding vasculature.

Historically, UGI followed by echocardiography and traditional angiography comprehensively evaluated for vascular ring/sling. Catheter angiography with rotational angiography and 3-dimensional reconstruction can provide exquisite
anatomic detail of the aorta and the great vessels (Figure 3); however, invasiveness of the study and presence of ionizing radiation limit the desire for its application. This combination of imaging studies as the standard workup has pre-dated the advent of high-quality cross-sectional angiography using either computed tomography (CT) or MRI, which tout up to 100% sensitivity for vascular ring evaluation. The ability to noninvasively visualize in detail the entire vascular ring or sling and the associated extravascular and extra-cardiac findings is a major benefit of using CT and/or MRI cross-sectional angiography over the aforementioned historic evaluation scheme.

When determining which modality to use, one must consider availability, risks and benefits. While CT angiography (CTA) carries risks associated with iodinated contrast administration and exposure to ionizing radiation, the modality may be more available than MRI, and requires little time for image acquisition. CT imaging enables accurate contrast bolus timing and decreases the need to sedate patients, which carries its own risks and would have a greater impact on a pediatric population as compared to adults. In a patient who cannot remain immobile for MRI and is not a suitable candidate for sedation or anesthesia, the rapid scan time of CT would be most beneficial.

Another strength of CTA is its high spatial and temporal resolution that produces clear visualization of the airways. In cases of suspected tracheobronchomalacia in association with vascular ring/sling, CTA may be the modality of choice. The use of prospective gating with CTA enables visualization of concomitant cardiac anomalies; however, this increases the radiation dose to the patient.

MRI protects the patient from risks associated with iodinated contrast administration and exposure to ionizing radiation, which imparts a cumulative risk of malignancy. MRI also allows for simultaneous multiplanar imaging of associated cardiac abnormalities and can be used to evaluate the trachea. Despite these benefits, MRI is not a risk-free modality for vascular imaging. MRI examinations for vascular rings take more time to perform than CT, increasing the need for sedation in pediatric patients. As MRI technology has advanced, rapid sequences combined with K-space
undersampling and parallel imaging techniques have shortened scan time and partially mitigated the need to sedate some patients. In many cases, contrast administration is unnecessary. In younger children, the vasculature may be best evaluated with contrast-enhanced magnetic resonance angiography (MRA); however, this exposes the patient to the risks of gadolinium-based contrast administration, including nephrogenic systemic fibrosis and cerebral gadolinium deposition.

Cross-sectional images can be used to produce 3-dimensional models of the great vessels using 3D printing. While still in its infancy, this technology may be increasingly used as a roadmap for surgeons performing corrective surgery.

**Classification**

Vascular rings are anomalies in which the trachea and esophagus are completely encircled by vascular tissue, although some portion of the vascular ring may be atretic. When a portion of the ring is atretic (e.g., a portion of a double aortic arch), the diagnosis of a vascular ring must be inferred from other imaging “clues,” as vascular imaging modalities depend on the flow of blood through an open lumen. The presence of one of the “3Ds” indicates a vascular ring in all cases when it occurs opposite the side of the aortic arch: Diverticulum, Dimple, or Descending aorta contralateral to the aortic arch (circumflex arch). A diverticulum is a dilated out-
Aortic Vascular Rings and Pulmonary Sling, Poletto

pouching off the descending aorta that gives rise to the aberrant contralateral subclavian artery. A dimple is a small blind pouch off the descending aorta.

The types of vascular rings may be classified as follows:

- double aortic arch
- right aortic arch
  - with diverticulum of Kommerell
  - with left descending aorta (right circumflex aortic arch)
  - mirror-image branching with retroesophageal left ductus
- left aortic arch
  - with diverticulum of Kommerell
  - with right descending aorta (left circumflex aortic arch).

**Double Aortic Arch**

Double aortic arch is the most common form of vascular ring, comprising approximately 50% to 60% of vascular rings. Typically, the right arch is more cephalad than the left. Both arches may be patent or one may be hypoplastic (usually the left) or atretic (also usually the left). In up to 73% of cases, the right arch will be dominant. The descending aorta can be on the left (most common), right or midline. Double aortic arch is most commonly an isolated finding and not associated with other congenital heart disease.

Chest radiography may show variable extrinsic mass effect on the right and (rarely) the left inferior trachea on the frontal view. A more reliable finding is anterior bowing of the inferior trachea on the lateral view. There will be extrinsic mass effect on the right and left aspects of the esophagus on frontal views during UGI, and extrinsic mass effect on the posterior esophagus on lateral views.

CT and MRI will show the dual aortic arches and their branch patterns. Classically, an axial image through the level of the branch arteries will show symmetry of the common carotid and subclavian arteries. As the vessels encircle the trachea, it will become narrowed. Volume-rendered images of the airway provide exquisite visualization of luminal narrowing. The proximal thoracic esophagus may be

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**FIGURE 9.** Lateral view from esophagram in a neonate with aberrant subclavian artery. There is abnormal extrinsic impression on the posterior aspect of the upper thoracic esophagus (arrow).

**FIGURE 10.** CT and MRI images in children with right arch with aberrant left subclavian arteries arising from diverticulum of Kommerell. On the axial CT image (A), there is dilation of the left subclavian artery where it arises from the proximal descending aorta, representing a diverticulum of Kommerell. Discrete narrowing at the junction of the diverticulum and the origin of the left subclavian artery (arrow) suggests the presence of a left-sided ligamentum arteriosum. Flattening of the posterior margin of the trachea also makes the presence of a vascular ring likely. Volume-rendered images from CT on the same patient (B) provide a 3-dimensional appreciation of the impact of the aorta on the airway. Coronal image from post-contrast MR angiography (C) in a different patient demonstrates the drastic change in caliber from the diverticulum of Kommerell to the origin of the subclavian artery (arrowhead).
dilated cephalad to the vascular ring. In cases where the arches are codominant, an advantage of MR is that phase contrast sequences can quantitate which arch has more flow.

Cases of right dominant double aortic arch with partially atretic left arch fall into two variants: atresia between the left common carotid and the left subclavian artery, and atresia distal to the left subclavian artery. In the former, the vascular ring can look identical to the right aortic arch with aberrant left subclavian artery from a diverticulum of Kommerell. Imaging may not be able to differentiate these two entities. While both are vascular rings, the surgeon must be prepared to ligate an atretic portion of the left arch in addition to a ligamentum arteriosum. In such cases, the subclavian arterial flow is reconstituted via retrograde flow down the vertebral artery (Figure 8).

Right Arch with Aberrant Subclavian Artery (with Diverticulum of Kommerell)

Right arch with aberrant subclavian artery arising from a diverticulum of Kommerell is the second most common vascular ring. It is not commonly associated with intracardiac defects, in contrast to the nonring right aortic arch with mirror image branching. The first arch vessel is the left common carotid artery, followed by the right common carotid artery, the right subclavian artery, and finally the left subclavian artery arising from the posterior aspect of the aortic arch. The left-sided ligamentum arteriosum completes the vascular ring. In such cases, the left subclavian artery arises from a diverticulum of Kommerell, the remnant of the left dorsal aorta. The left-sided ligamentum arteriosum typically inserts at the junction of the diverticulum and the left subclavian artery. In up to 10% of patients with a right arch and aberrant subclavian artery, the ligamentum arteriosum is right-sided and the diverticulum of Kommerell is absent, indicating no vascular ring.

Cases with atretic left arch distal to the left subclavian artery can look similar to right aortic arch with mirror image branching. What differentiates these entities is the course of the first vessel arising from the aortic arch on axial imaging. In mirror image branching, the left innominate artery courses relatively anteriorly, while in double aortic arch with partially atretic left arch, the left arch will course posteriorly (Figure 7), allowing encirclement of the trachea. In addition, double aortic arch with a partially atretic left arch will have a dimple on the descending aorta, which represents the remnant of the left dorsal aorta; mirror image branching does not have a dimple. Rarely the left aortic arch is atretic at the origin of the left subclavian artery. In such cases, the subclavian arterial flow is reconstituted via retrograde flow down the vertebral artery (Figure 8).
anterior bowing of the inferior trachea on the lateral view. The appearance is typically indistinguishable from that of a double aortic arch. On UGI, the right arch will have extrinsic mass effect on the right aspect of the esophagus on frontal views, and the diverticulum of Kommerell will cause extrinsic mass effect on the posterior esophagus on lateral views. The posterior impression from the diverticulum will typically be smaller than that of double aortic arch or circumflex arch (Figure 9).

CT and MRI best distinguish right arch with aberrant left subclavian artery as a vascular ring from those that are not. The ligamentum arteriosum is visible if it is partially patent or calcified; this is better seen on CT than MRI. The presence of a left-sided ligamentum arteriosum, diverticulum of Kommerell, and tracheal narrowing are all suggestive of a vascular ring (Figure 10). If the proximal aberrant subclavian artery maintains uniform caliber, and there is no airway narrowing, then a vascular ring is unlikely.

When the diagnosis of vascular ring is questionable, the presence/absence of symptoms may guide decisions to perform corrective surgery.

**Right Aortic Arch with Left Thoracic Descending Aorta (Circumflex Aortic Arch)**

In the third most common type of vascular ring, a right aortic arch crosses midline in the superior thorax, coursing posterior to the trachea and esophagus, then descends to the left of midline. The branching pattern may have a left innominate artery or an aberrant left subclavian artery. In either case, there is a vascular ring when there is a left-sided ligamentum.

On radiography, the vascular ring is inferred by the presence of a left descending aorta opposite a right-sided arch and anterior bowing of the trachea. As with other vascular rings, there will be an abnormal impression on the posterior esophagus during esophagram. The size of the impression will be similar to that of a double aortic arch. When there is only a single right-sided aortic impression on the esophagus on the frontal view and a large posterior impression on the lateral view, the diagnosis of circumflex aorta is suggested (Figure 11).

As with other vascular rings, CT and MRI will provide definitive diagnosis of the circumflex arch, delineate the branch pattern, and assess the effect of the vessels on the airway, allowing for surgical planning (Figure 12).

**Right Aortic Arch with Retroesophageal Left Ductus**

The diagnosis of this rare type of vascular ring may be challenging as its branching pattern is identical to the nonring mirror image right aortic arch. In right aortic arch with mirror image branching, the ductus may be: 1) right-sided from the aortic arch, 2) left-sided off the base of the innominate artery, or 3) left-sided from the right descending aorta (vascular ring). In the last case,
the retroesophageal ligamentum completes the ring after the ductus closes. The presence of a subtle “dimple” on the right descending aorta, directed toward the left, indicates the presence of the retroesophageal ligamentum and signifies a vascular ring.

**Vascular Rings with Left Arch**

Vascular rings in the setting of left aortic arch are very rare. Although much less common than the right aortic arch variant, left aortic arch with retroesophageal diverticulum was the vascular ring first described by Kommerell on barium esophagram in 1936.\(^1\) This anomaly is the mirror image of right arch type, with the vascular ring completed by a right-sided ligamentum. Similarly, left aortic arch with right descending aorta (left circumflex aortic arch) is a mirror image of the right-sided lesion with the ring completed if there is a right-sided ligamentum (Figure 13).

**Pulmonary Sling**

Pulmonary sling is exceedingly rare; a Taiwanese study of a population of over 100,000 children found an incidence of 1:17,000 on screening echocardiogram.\(^18\)

Diagnosis of pulmonary sling can rarely be made from radiography alone. Asymmetric aeration of the lungs on the frontal view with indentation of the posterior distal trachea on the lateral view suggests a pulmonary sling, with the smaller lung supplied by the pulmonary artery of abnormal origin. The mainstem bronchi may be displaced inferiorly and have a horizontal course, with an inverted T-shaped appearance.\(^14\) The anomalous left pulmonary artery may obstruct the right mainstem bronchus. In such cases, the right lung may have retained fetal fluid at birth and then become progressively hyperlucent.

As this is the only anomalous vessel to course between the trachea and esophagus, on UGI there will be an abnormal impression on the anterior esophagus, and the trachea will be displaced.

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**FIGURE 15.** Tc99m-MAA perfusion scan in the same child as Figure 13. There is global decreased perfusion to the left lung with only 38% of pulmonary arterial blood going to the left lung.
anteriorly. Fluoroscopy can also be used to dynamically evaluate the airway for tracheomalacia.

As with vascular rings, CT and MRI will definitively make the diagnosis, and can assess the impact of the pulmonary vessels on the airway (Figure 14). As most of these patients have complete tracheal rings, the trachea may have a round configuration on axial imaging and may be relatively small in caliber. Other airway anomalies can occur, most commonly a bridging bronchus (a bronchus arising from a left- or right-sided bronchus, crossing the midline, supplying a portion of the contralateral lung). Such small airway abnormalities are better seen on CT than MRI.

An advantage of MRI is that it can quantitate arterial perfusion of each lung; perfusion of the left lung may be decreased. Cine MRI can also be used to dynamically evaluate the trachea throughout the respiratory cycle. Pulmonary perfusion can also be assessed with nuclear lung perfusion, utilizing technetium 99m-labeled microaggregated albumin (MAA) (Figure 15).

**Repair of Vascular Rings and Pulmonary Sling**

For repair of vascular rings, left lateral/posterirolateral thoracotomy is used. The surgeon will run a finger along the esophagus to ensure the ring component is present. In cases where the left component of the vascular ring is an atretic arch or a ligamentum arteriosum, the proximal and distal ends are divided. Usually the tissue is under tension and a gap of 2–3 cm will occur after the tissue is divided, which should suffice in loosening the vascular ring.

In cases of double aortic arch, the smaller of the two arches is ligated. After proximal and distal control of the smaller arch are obtained, a test occlusion is performed and the blood pressure is assessed to ensure that no pressure gradient occurs. After dividing, the two ends are oversewn. Again, the area is usually under tension and a gap will occur.

In our practice, we begin and end each case with bronchoscopy, and assess the airway for airway compromise. Any patient with significant residual compression is followed by our pulmonologists. Long-term outcome for these patients is good, with approximately 5% of patients requiring re-repair for recurrent symptoms.

The approach for a pulmonary artery sling is anteriorly utilizing a median sternotomy. The left pulmonary artery is reimplanted on the main pulmonary artery, anterior to the trachea. An excessively long or redundant left pulmonary artery may need to be partially resected to prevent kinking. Prior compression of the airway causes a longitudinal stenosis, which may require a sliding-plasty of the trachea. The patient may require cardiopulmonary bypass during the resection of the stenotic segment of the trachea, then a sliding of the posterior and anterior segments of the trachea for reconstruction. The reconstructed trachea will then need time to heal; monitoring by a pulmonologist or otolaryngologist is appropriate.

**Summary**

Vascular rings and pulmonary sling are a diverse group of congenital thoracic vascular anomalies. Patients present from the prenatal period into adulthood, depending on the extent of airway and esophageal compression. While radiographs and fluoroscopy are excellent screening tools, they poorly distinguish the types of vascular rings. Echocardiography provides excellent anatomic detail, but detailed evaluation is not always possible and airways cannot be evaluated. Cross-sectional imaging (CT and MRI) have become the gold standard in evaluation since they can completely evaluate vascular and airway anatomy, obviating the need for catheter angiography. The risks and benefits of CT and MRI should be considered prior to imaging. Surgical correction has excellent long-term success.

**REFERENCES**


Acute chest pain presents a clinical challenge because of its prevalence, broad differential, and risk of serious morbidity and mortality. The diagnosis is complicated by its spectrum of presentations, including the most severe ST-elevation myocardial infarction (MI) and unstable angina (UA), where biomarker evidence of myocardial damage is lacking. Event prediction remains difficult even if coronary atherosclerotic disease (CAD) can be demonstrated, as the majority of plaque ruptures are—perhaps surprisingly—clinically silent, with acute coronary syndromes (ACS) occurring stochastically in proportion to CAD burden. If patients can be determined to be CAD-negative, however, they have essentially zero risk, both in the short and long term.

This review discusses the state of coronary CT angiography (CCTA) for management of acute chest pain in the emergency department (ED). CCTA directly visualizes coronary plaque burden, thereby ruling out a greater proportion of negative patients than any other noninvasive test. Simultaneously, it delivers the best short-term diagnostic accuracy in comparison to existing accelerated diagnostic protocols (Table 1). CCTA also provides the best long-term prognosis at the earliest time point, as well as positive effects on downstream morbidity and mortality. Nevertheless, the present level of CCTA utilization does not reflect its superior ability. We will explore the effectiveness of CCTA and advocate for its intelligent use in this patient population.

Non-CCTA Accelerated Diagnostic Protocols

Chest pain places a significant burden on the ED and the health care system as a whole. Standards of care include clinical observation, serial electrocardiograms (ECGs), serial cardiac biomarkers, and provocative functional or imaging tests, with the overall goal of reducing short-term major adverse cardiovascular events (MACE). Several accelerated diagnostic protocols have been developed to reduce ED and total costs without compromising patient safety.

The TIMI (Thrombolysis In Myocardial Infarction) score was initially developed to assess ACS severity in diagnosed patients; it has been used with varying degrees of success to predict ACS itself. The ADAPT accelerated diagnostic pathway combines the lowest possible TIMI score of zero with negative conventional troponin-I at 2 hours to reduce the risk of ACS to 0.3%. The ADAPT pathway has a specificity of approximately 25%, meaning only approximately a quarter of disease negative patients actually test negative. The remaining three quarters test positive, are not “ruled out,” and are exposed to additional testing. A randomized clinical trial (RCT) employing ADAPT vs. standard of care found that ADAPT doubles the early discharge rate without compromising patient safety. The APACE pathway uses high-sensitivity troponin-I at 2 hours with a TIMI score as high as 1 to achieve essentially the same low-risk rate as ADAPT and a specificity proportion of close to 50%.

The HEART risk score was specifically constructed to predict the risk of ACS and was validated within the ED patient population. The HEART pathway successfully combines a low-risk HEART score with a 3-hour conventional troponin-I to reduce the risk of MACE rate to zero with a specificity proportion of approximately 50%. ADAPT and APACE recommend outpatient functional testing following early discharge due to a fatality within the study cohort in which a stress test was performed but incorrectly interpreted. Notably, the HEART pathway investigators suggest that based on their results, low-risk patients require no further outpatient testing. Additional protocols have been suggested (Table 1).

Acute setting accelerated diagnostic pathways must avoid provocative testing because of the real—albeit unlikely—possibility of provoking infarction. Functional accelerated protocols must therefore rely on the resting state as a pseudo-stress equivalent. Acute
rest single-photon emission computed tomography (SPECT) myocardial perfusion imaging (MPI) demonstrates a 99% negative predictive value (NPV) for acute MI, but examinations must be read critically with a resultant loss of specificity.\textsuperscript{10,11} Moreover, these findings are for MI only. With respect to all short-term MACE, the miss rate appears closer to 7%.\textsuperscript{10} In practice, an RCT employing SPECT after negative biomarker testing demonstrated reduced admissions with no change in short-term outcome. However, widespread implementation of this protocol is unlikely due to the practical difficulties surrounding the unscheduled use of radionuclide pharmaceuticals.\textsuperscript{12}

### CTTA-based Acute Chest Pain Management

CCTA for patients with acute chest pain is more effective at many levels. A systematic review and meta-analysis performed in 2008 yielded an NPV of 100% and specificity of 89% in predicting significant CAD.\textsuperscript{13} In 2012, a systematic review and meta-analysis concluded that CCTA has an NPV of 99.3% for ACS and a specificity of 87%, meaning only approximately 1 in 10 negative patients are exposed to further testing.\textsuperscript{14} Three ensuing large multicenter RCTs compared CCTA and functional protocols and demonstrated that CCTA accelerates diagnosis, with twice to quadruple as many direct discharges from the ED, a reduction in ED costs of 18% to 38%, and no increase in short-term MACE.\textsuperscript{15} An RCT comparing CCTA with the cheapest and most widely used functional test, treadmill exercise stress ECG, shows that costs with CCTA are still lower, primarily driven by decreased length of stay.\textsuperscript{16} In 2014, the American Heart Association-American College of Cardiology Non-ST Elevation Acute Coronary Syndrome (AHA/ACC NSTE-ACS) guidelines assigned CCTA the highest level of evidence. The 2015 AHA/ACC released appropriate use criteria jointly with the American College of Radiology (ACR) that endorse CCTA as a first-line exam.\textsuperscript{17}

### Table 1. Test characteristics for ED chest pain management strategies in terms of short-term MACE, assuming population with 15% prevalence

<table>
<thead>
<tr>
<th>Management strategy</th>
<th>Positive Likelihood Ratio</th>
<th>Negative Likelihood Ratio</th>
<th>Risk if Protocol Positive (%)</th>
<th>Risk if Protocol Negative (%)</th>
<th>Share of Negative Patients Ruled Out (%)</th>
<th>Share of Positive Patients Ruled In (%)</th>
<th>Overall Diagnostic Accuracy (%)</th>
<th>Level of Evidence</th>
</tr>
</thead>
<tbody>
<tr>
<td>Clinical judgement</td>
<td>1.32</td>
<td>0.14</td>
<td>19</td>
<td>2.5</td>
<td>27.4</td>
<td>96.1</td>
<td>85.8</td>
<td>Prospective multicenter study\textsuperscript{42}</td>
</tr>
<tr>
<td>TIMI score = 0</td>
<td>1.30</td>
<td>0.11</td>
<td>19</td>
<td>1.9</td>
<td>25.0</td>
<td>97.2</td>
<td>86.4</td>
<td>Retrospective meta-analysis\textsuperscript{43}</td>
</tr>
<tr>
<td>TIMI score = 0 and 2-hour conventional troponin-I (ADAPT Pathway)</td>
<td>1.30</td>
<td>0.01</td>
<td>19</td>
<td>0.2</td>
<td>23.4</td>
<td>99.7</td>
<td>88.3</td>
<td>Prospective multicenter study\textsuperscript{44} and single-center RCT \textsuperscript{13}</td>
</tr>
<tr>
<td>TIMI score ≤ 1 and 2-hour high-sensitivity troponin-I (APACE Pathway)</td>
<td>1.86</td>
<td>0.01</td>
<td>25</td>
<td>0.2</td>
<td>46.5</td>
<td>99.4</td>
<td>91.5</td>
<td>Prospective multicenter study\textsuperscript{45}</td>
</tr>
<tr>
<td>Low-risk HEART score</td>
<td>1.69</td>
<td>0.09</td>
<td>23</td>
<td>1.5</td>
<td>43.1</td>
<td>96.3</td>
<td>88.3</td>
<td>Prospective multicenter study\textsuperscript{46}</td>
</tr>
<tr>
<td>Low-risk HEART score and 3-hour conventional troponin-I (HEART Pathway)</td>
<td>1.98</td>
<td>0.00</td>
<td>26</td>
<td>0.0</td>
<td>49.6</td>
<td>100.0</td>
<td>92.4</td>
<td>Single-center RCT \textsuperscript{8}</td>
</tr>
<tr>
<td>Presentation high-sensitivity troponin-T and clinical gestalt</td>
<td>2.04</td>
<td>0.06</td>
<td>26</td>
<td>1.0</td>
<td>52.5</td>
<td>97.0</td>
<td>90.3</td>
<td>Prospective single-center study\textsuperscript{47}</td>
</tr>
<tr>
<td>Presentation high-sensitivity troponin-I</td>
<td>2.29</td>
<td>0.02</td>
<td>29</td>
<td>0.3</td>
<td>56.8</td>
<td>98.9</td>
<td>92.6</td>
<td>Prospective multicenter study\textsuperscript{48}</td>
</tr>
<tr>
<td>Acute rest SPECT MPI alone</td>
<td>2.08</td>
<td>0.38</td>
<td>27</td>
<td>6.3</td>
<td>63.5</td>
<td>75.8</td>
<td>74.0</td>
<td>Prospective multicenter study\textsuperscript{15}</td>
</tr>
<tr>
<td>Acute CCTA alone</td>
<td>7.31</td>
<td>0.06</td>
<td>56</td>
<td>1.0</td>
<td>87</td>
<td>95</td>
<td>93.8</td>
<td>Systematic review and meta-analysis of smaller studies (one RCT, 5 prospective and 3 observational cohorts)\textsuperscript{14}</td>
</tr>
</tbody>
</table>
The benefit of CCTA continues in the high-sensitivity troponin era, although its advantages have recently been questioned. A 2016 RCT in the Netherlands comparing CCTA to usual care including high-sensitivity troponins showed a 6% increase in direct ED discharges with CCTA, 65% vs. 59%, although without attaining statistical significance. The authors suggest their study was underpowered to show the small CCTA ED discharge rate benefit. They also note that the integrated nature of the Netherlands health system and excellent access to primary care steered a greater proportion of low-risk, CAD-negative and, hence, CCTA-negative patients away from the ED. The diagnostic power of CCTA rests in its excellent ability to identify patients without CAD and, therefore, without risk; thus, as CAD burden increases within the test population, the diagnostic accuracy decreases. From RCTs in the United States, it has been calculated that cost savings will occur only in populations where CCTA will show < 50% stenosis in at least 72% of patients, ie, populations with relatively lower risk of ACS. The Netherlands study just barely met this quota, with 74% of patients negative for obstructive CAD. These caveats suggest that in a U.S. population with lower risk, the ED discharge rate for a combined CCTA and higher-sensitivity troponin strategy might be higher.

Moreover, although the Netherlands study reports that both cohorts demonstrated similar median lengths of stay, 6.3 hours, outcomes within the next quartile of patients dramatically differed. Specifically, 75% of CCTA cohort stays were < 11.1 hours, while 75% of patients within the usual care group were not discharged until >25.5 hours. CCTA also prevented additional downstream testing. It is not surprising that CCTA led to statistically significant lower short-term costs, a savings of approximately one-third, despite the higher prevalence of CAD. The different findings from the U.S. studies may reflect a practice pattern emphasizing earlier ED disposition decisions, but certainly do not reduce the central conclusion of shorter stays and lower costs using CCTA.

### Technical Improvements in CCTA

The spatial resolution of CCTA is typically 0.35 mm as compared to 0.16 mm for invasive angiography. Whereas a 3-mm coronary lumen is delineated on 18 pixels in fluoroscopy, CCTA displays the same vessel over 9 voxels, limiting determination of the exact degree of CAD. CCTA stenosis severity is therefore reported in increments of 25%. While interventional coronary angiography (ICA) outpaces CCTA in differentiating patients with varying degrees of CAD severity, surpassing CCTA’s positive predictive value (PPV), the NPV of CCTA at least equals that of ICA.

CCTA continues to improve technically. Traditionally, CCTA struggled to match the temporal resolution of the invasive exam. Fluoroscopy yields 30 frames per second, corresponding to a resolution of 33 milliseconds, essentially eliminating motion artifact. Increased CCTA temporal resolution on an ECG-gated exam is now achieved using multicycle reconstructions, higher gantry rotation speeds, use of multiple x-ray sources, and wider detectors.

Radiation dose in CCTA initially matched the effective dose of SPECT...
MPI exams of approximately 12-14 mSv; however, prospective gating, faster scanning, and better reconstruction algorithms have halved the dose, with improvement continuing. By comparison, a noncomplicated diagnostic cardiac catheterization delivers 8-10 mSv. Controversy exists as to whether exposure < 50 mSv imparts any increased risk, with major societies at odds about appropriate recommendations. Assuming, for the sake of caution—as the ACR does—the absence of a threshold for radiation-induced damage, the lifetime attributable risk of fatal cancer for a 5 mSv study would be a single additional fatal cancer per 2,000 examinations, undoubtedly a small fraction of the likely study benefit. The latest CT scanners deliver diagnostic scans at approximately 1 mSv.20,23 At such doses, radiation is no longer a realistic concern.

A principal technical advantage of CCTA is that the data obtained is intrinsically 3-dimensional. Images from a single acquisition can be viewed from any angle or projection with no vessel overlap, unlike the 2-dimensional data produced by angiography. This benefit partially compensates for CCTA’s lesser spatial and temporal resolution. Commonly used projections and reformations include surface rendering (Figure 1), curved multiplanar reconstructions (Figure 2), and straightened multiplanar reconstructions along the course of the vasculature (Figure 3).

High-risk Coronary Features Indicate Increased Risk

CCTA demonstrates prognostically important pathology not seen on catheter angiography. High-risk CCTA coronary plaque features include positive remodeling (increase in the outside diameter of the vessel, Figure 4), spotty calcium (Figure 5A), low attenuation plaque (Figures 5A-B), and rim-enhancing plaque (the “napkin-ring” sign, Figure 6), the latter likely indicating the presence of a lipid-laden plaque core. The relative risk of these high-risk plaque features for short-term MACE within the ED population is comparable to that of obstructive CAD, approximately 30 times. This holds true even after controlling for the presence of obstructive disease, thus increasing exam utility. CCTA-positive patients with nonobstructive CAD and without high-risk features could be classified as lower risk, increasing the proportion of patients that CCTA could clear. In patients with obstructive CAD and high-risk features, the PPV of the exam increases.24 High-risk features also serve as a biomarker of functional disease, as both plaque volume and high-risk characteristics correlate with the functional significance of a lesion as determined by invasive measurement of the pressure differential across the lesion, ie, ICA with measurement of the fractional flow reserve (FFR).25

Long-term Prognosis: The 6-Year CCTA Guarantee

In outpatients, a completely negative CCTA provides a virtual guarantee of approximately 6 years of MACE-free survival, while nonobstructive CAD indicates a stable small risk for at least 2 years.26 The risk of MACE is approximately 0.5% per year for limited nonobstructive CAD and between 2-3 times that number with obstructive disease or extensive non-obstructive disease.1 Risk increases with the number of vessels involved. Just as high-risk features affect short-term risk, they also increase risk over the long term.26,27

No stress modality can rival CCTA in detecting nonobstructive disease, since stress testing is blind to disease that is not flow limiting. Thus, only CCTA can provide information about the early CAD stages noninvasively. For this same reason, CCTA provides a longer guarantee if negative, in addition to comparable prognostic information when positive. Comparison between CCTA and exercise ECG actually demonstrates increased long-term risk in CCTA-positive patients irrespective of functional testing results.28 CCTA and SPECT can work synergistically to risk-stratify patients at the cost of increased radiation dose.27
gests that revascularization should be
stress tests at approximately 70% vs.
respectively. The relative risk of a false-positive
CCTA after a functional test, for exam-
clinical benefits of CT-FFR compared to
invasive FFR as the reference standard
validates its accuracy; CT-FFR improves
both the sensitivity and specificity of the
CCTA exam.35 However, the PLAT-
FROM RCT showed no downstream
clinical benefits of CT-FFR compared to
CCTA.36,37 Although the rate of future
clinical deterioration may depend on the
functional deficit, it likely primarily flows
from the background clinical and anatomi-
al risk, especially if assessed earlier
and/or in a lower-risk population.
Finally, the CATCH trial directly elu-
cidates the long-term impact of CCTA
use within the ED population, despite
the difficulty in assembling enough pa-
ents and events to power a study of this
kind.38 In CATCH, patients with poten-
tial ACS admitted for less than 24 hours
underwent both functional testing and
CCTA prior to discharge, but the CCTA
data was reported to the referring physi-
cian in only half of the patients. After 18
months, the CCTA cohort demonstrated
a 2% reduction in cardiac death or MI.
This difference was just shy of statistical
significance, with p-value of 0.06. The
usual care cohort, in contrast, had a
statistically significant increased risk of all
MACE, 5% vs. 2%, and when including
readmission, had a statistically signif-
icant higher risk of adverse outcomes,
16% to 11%. Complementing these
findings, retrospective analysis of CCTA
patients shows five-fold lower odds of
recidivism.39 These results demonstrate
a morbidity and mortality benefit to early

Clinical Benefit to Anatomical over
Functional Testing

Long-term studies reveal a clinical
benefit to CCTA. Not surprisingly,
CCTA outperforms functional tests
when anatomical findings on ICA are
used as the reference standard. Within a
population of 6200 patients undergo-
ing CCTA after a functional test, for ex-
ample, the relative risk of a false-positive
or false-negative stress test as com-
pared to an inaccurate CCTA was 1.4 and 3.1,
respectively.29 An analysis of the nation-
wide elective ICA registry confirms the
higher PPV of CCTA vs. all the various
stress tests at approximately 70% vs.
46%.30 Newer research, however, sug-
gests that revascularization should be
guided by the functional perfusion deficit
as measured by FFR during ICA, and a
systematic review and meta-analysis
demonstrate that SPECT outperforms
CCTA in identifying lesions when ICA
with FFR is used as the new gold stan-
dard.31 One would expect, based on this
research, to find clinical benefit for non-
invasive functional testing; however, the
opposite has been shown.
A meta-analysis of recent clinical
trials in outpatients with CAD (PROM-
ISE, SCOT-HEART, and two smaller
RCTs) shows a statistically significant
reduction of annual MI in patients who
underwent CCTA, to approximately
0.7 times the rate of patients with func-
tional exams.32 Three-year follow-up of
SCOT-HEART similarly demonstrates a
statistically significant cardiac fatality
and MI risk ratio of 0.5. This benefit is
attributed to statistically significant
changes in medical and invasive man-
agement.33 CCTA is already a first-line
test for outpatient CAD in Europe, and
these results suggest it should be first-
line in the United States as well.34
Undoubtedly, CCTA could be im-
proved if functional data could be derived
from the exam. CT-FFR is a computa-
tional technique that noninvasively cal-
culates the pressure gradient across lesions
using the 3-dimensional coronary anat-
omy, without catheterization, without
adenosine administration, and with no
increase in contrast material injected or
in radiation dose. A meta-analysis with
invasive FFR as the reference standard
validates its accuracy; CT-FFR improves
both the sensitivity and specificity of the
CCTA exam.35 However, the PLAT-
FORM RCT showed no downstream
clinical benefits of CT-FFR compared to
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clinical deterioration may depend on the
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16% to 11%. Complementing these
findings, retrospective analysis of CCTA
patients shows five-fold lower odds of
recidivism.39 These results demonstrate
a morbidity and mortality benefit to early

FIGURE 3. Straightened multiplanar reconstruction of normal left main and anterior descending coronary arteries. The vessels must be completely assessed in 2 orthogonal projections. The bottom panel demonstrates a longitudinal view of the straightened vessels, while the top 5 panels demon-
strate selected cross-sectional slices corresponding to the colored vertical lines on the straight view. The axis for the longitudinal view is indicated by the red central dots within the top panels. LMCA = left main coronary artery; LCx = left circumflex artery; LAD = left anterior descending artery.

FIGURE 4. Positive remodeling. Longitudinal view (A) demonstrates a calcified plaque sitting dis-
tal to the trifurcation of the left main coronary artery and opposite the takeoff of the first di-
agonal. The lumen of the coronary artery is not narrowed, but the outside diameter of the vessel is increased, indicative of positive remodeling (PR). Cross-sectional view (B) re-demonstrates the intact coronary lumen.
use of CCTA in the ED population, just as in outpatients. In the long term, CCTA leads to an increase in downstream procedures, which degrades the initial cost-savings. However, a meta-analysis of the ED RCTs, which suggests a relatively matched increase of 2% in the patients who undergo ICA and revascularization after CCTA, also suggest that these major procedures are performed appropriately. These increased revascularizations may drive the clinical benefits discussed above. Impressively, retrospective analysis found a seven-fold lower likelihood of undergoing ICA without revascularization in ED patients triaged by CCTA. ED providers and radiologists should be aware that patients who receive early testing with CCTA rather than downstream functional tests are well-served clinically.

CCTA includes a final ancillary benefit: The most common incidental finding on CCTA is a pulmonary nodule, occurring at approximately the same rate as the general population, one in five. One in 10 nodules may represent a neoplasm, with a potential morbidity and mortality benefit of 1% if just half can be detected. Further work is necessary, but recent studies suggest additional downstream costs are minimal.

**Suggestions for Management**

The CAD-RADS (Reporting and Data System) has been introduced to standardize CCTA reporting and guide further research and management. Highlights are as follows: Patients with left main coronary stenosis of ≥50%, any vessel stenosis of 70%, or 3-vessel disease are described as CAD-RADS-4 and may be candidates for revascularization. In the acute setting, this score indicates that ACS is at least likely. CAD-RADS-3 includes any other stenosis >50% where ACS is possible and where admission and at least a functional test for further risk stratification would be considered appropriate. CAD-RADS-2 includes stenoses between 25% and 49%, where ACS is unlikely despite the presence of CAD, although high-risk plaque features would lead to a CAD-RADS-2/V designation and should escalate level of concern and further care.

**Conclusion**

CCTA for acute chest pain provides powerful diagnostic and prognostic information, decreases ED length of stay, reduces ED costs, and provides tangible clinical benefits. The latest biomarkers do not alter these findings. Accelerated diagnostic pathways that encourage early discharge without objective anatomic testing deny patients the proven benefits of CCTA. Further trials will hopefully lead to varied high-sensitivity troponins or integrated risk score cutoffs that stratify patients even better. Until that time, the use of early CCTA to further guide care represents the evidence-based strategy of maximal benefit and least harm, as is strongly supported by data.

**REFERENCES**


Mitral Annular Mass

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1 Department of Radiology and 2 Department of Medicine/Cardiology, Jacobi Medical Center, Albert Einstein College of Medicine, Bronx, NY

Case Presentation

A 74-year-old man presented with a multifocal acute cerebrovascular accident. Laboratory data revealed normal renal function and serum calcium level. Chest radiograph demonstrated an enlarged cardiac silhouette, but was otherwise unremarkable for age. A cardiac source for the emboli was suspected, and an echocardiogram was requested. Echocardiogram showed an intracardiac mass (Figure 1A), which was further investigated with computed tomography (CT), (Figure 1B, 1C) and MRI (Figure 1D).

FIGURE 1. Two-dimensional echocardiogram apical four-chamber view (A) shows a lobulated echogenic 3 × 2-cm mass in the region of the posterior mitral valve leaflet (arrow) that was mobile. Unenhanced CT four-chamber view image (B) shows a rim-calcified mass (arrow) corresponding to the echocardiogram abnormality with adjacent sheet-like calcifications (arrow head) that are likely in the mitral valve leaflet. EKG-gated enhanced CT maximum intensity projection (MIP) image in short axis projection though the mitral valve (C) reveals a centrally hypodense, rim-calcified mass (white arrow) in the atrioventricular groove with close relationship to the posterior mitral valve leaflet. Calcifications of the mass are contiguous with the densely calcified inferior mitral annulus (arrowhead). The anterior mitral valve leaflet (black arrow) exhibits degenerative calcifications. EKG-gated cardiac MRI fast imaging employing steady-state acquisition (FIESTA) short axis image (D) through the same level as image C shows that the central content of the mass (arrows) has high-signal intensity surrounded by low-signal rim.
Key imaging finding(s)

Lobulated mass contiguous with the mitral annulus and posterior leaflet (CT and MRI).

Differential Diagnoses

Caseous calcification of the mitral annulus
- Myxoma
- Mitral para-annular blood cyst
- Mitral annular abscess
- Infective endocarditis

Caseous Calcification of the Mitral Annulus

Caseous calcification of the mitral annulus (CCMA) accounts for < 1% of mitral annular calcification (MAC) cases. It most commonly occurs in elderly women; however, it may also affect younger patients with advanced renal disease or other metabolic disorders that result in abnormal calcium metabolism. A calcified rim surrounds caseous or toothpaste-like material composed of calcium, fatty acids, and cholesterol, typically located on the posterior mitral annulus. On transthoracic echocardiogram (TTE), CCMA usually appears as a large, round, echogenic mass with smooth borders and central echolucency. On cardiac MRI, the mass most commonly has low-signal intensity on T1- and T2-weighted sequences. On CT, it typically appears as a round, smooth mass with central hyperdensity and peripheral dense calcifications. Due to the variable appearance of caseous calcifications as a result of liquefactive necrosis on imaging, the central component of the mass may less frequently appear hypoechoic or heterogeneous on MR sequences, as is seen in the illustrative case. Nevertheless, the densely calcified rim consistently appears hyperdense on CT and hypointense on MR. The lack of soft tissue attenuation and enhancement is a clue to differentiate CCMA from a tumor. Because misdiagnoses and unnecessary surgical treatment may occur, a combination of multiple imaging modalities is often necessary to establish or suggest the diagnosis. Microscopic examination shows an amorphous, acellular, basophilic, and calcific structure, with chronic inflammatory macrophages. CCMA is usually a benign and asymptomatic lesion with no need for therapeutic intervention. Rarely, if mitral dysfunction or conduction disturbance occurs, mass excision and valve repair or replacement is the preferred treatment. Cases of systemic embolization by CCMA have been reported, which likely explains the symptoms in our patient, and prompt intervention. CCMA usually is a benign and asymptomatic lesion with no need for therapeutic intervention. Rarely, if mitral dysfunction or conduction disturbance occurs, mass excision and valve repair or replacement is the preferred treatment. Cases of systemic embolization by CCMA have been reported, which likely explains the symptoms in our patient, and prompt intervention.

Myxoma

Cardiac myxoma represents the most common primary cardiac tumor in adults. It is typically a polypoid, intracavitary left atrial mass that arises from the interatrial septum, but it may originate in any cardiac chamber. Most affected patients present with at least one feature of a classically described triad that includes cardiac obstructive symptoms, constitutional symptoms, and embolic events. Echocardiogram features include an echogenic round, lobular, mobile mass, which seldom originates from the valves. Common nonspecific radiographic features include cardiomegaly and intracardiac calcification. Myxomas may cause elevated left atrial pressures with associated left atrial enlargement, vascular redistribution, a prominent pulmonary trunk, pulmonary edema, pleural effusions, and rarely an enlarged left atrial appendage.

On echocardiogram, the mass most commonly has low-signal intensity on T1- and T2-weighted sequences. On CT, it typically appears as a smooth mass with central hyperdensity and peripheral dense calcifications. Due to the variable appearance of caseous calcifications as a result of liquefactive necrosis on imaging, the central component of the mass may less frequently appear hypoechoic or heterogeneous on MR sequences, as is seen in the illustrative case. Nevertheless, the densely calcified rim consistently appears hyperdense on CT and hypointense on MR. The lack of soft tissue attenuation and enhancement is a clue to differentiate CCMA from a tumor. Because misdiagnoses and unnecessary surgical treatment may occur, a combination of multiple imaging modalities is often necessary to establish or suggest the diagnosis. Microscopic examination shows an amorphous, acellular, basophilic, and calcific structure, with chronic inflammatory macrophages. CCMA is usually a benign and asymptomatic lesion with no need for therapeutic intervention. Rarely, if mitral dysfunction or conduction disturbance occurs, mass excision and valve repair or replacement is the preferred treatment. Cases of systemic embolization by CCMA have been reported, which likely explains the symptoms in our patient, and prompt intervention. CCMA usually is a benign and asymptomatic lesion with no need for therapeutic intervention. Rarely, if mitral dysfunction or conduction disturbance occurs, mass excision and valve repair or replacement is the preferred treatment. Cases of systemic embolization by CCMA have been reported, which likely explains the symptoms in our patient, and prompt intervention.

Mitral Para-annular Blood Cyst

Congenital blood cysts of the heart valves most often involving the tricuspid and mitral valves of fetuses and infants are generally considered of no clinical significance. However, lesions may persist and enlarge to form giant cysts of the heart valves. Even when large, blood cysts are usually asymptomatic in adults and are often discovered incidentally during routine echocardiographic evaluation. In rare cases, mitral regurgitation can occur due to the para-annular location of the cyst. Cysts may be a potential source of cerebrovascular embolism. There is not necessarily a correlation between the size of the cyst and hemodynamic consequences, and there is no consensus regarding the management of blood cysts. Some authors suggest that asymptomatic cysts, because of their benign character, can be monitored with echocardiography, and that resection should be reserved for cysts that interfere with normal cardiac function. On echocardiogram, blood cysts appear as hypoechoic cystic lesions, which may contain internal echoes, often with smooth outer surfaces. On CT, they appear as hypodense cystic masses with or without wall calcification. On MRI, blood cysts demonstrate low to intermediate signal intensity on T1-weighted images and high signal intensity on T2-weighted images without enhancement.
Mitral Annular Abscess

Mitral annular abscesses are most often seen in patients who have had endocarditis or bacteremia. The patients typically present with fever, pain, fatigue and shortness of breath. The lesions usually appear along the mitral-aortic fibrosa. They may closely resemble CCMA on imaging, but lack calcifications, typically have a homogeneous core, and may show systolic blood flow by color Doppler. On echocardiogram, myocardial abscesses appear as hypochoic masses. Hypokinesis of the surrounding myocardium can be present, along with pericardial effusion. CT or MRI can be used to accurately define the anatomic location, as needed for surgical intervention. Abscesses appear on CT scan as loculated lesions with fluid density and peripheral contrast enhancement with surrounding myocardial wall thickening. MRI is more sensitive for abscess detection than other imaging modalities due to its superior temporal and spatial resolution; in addition, its use is advantageous for morphological evaluation, since it provides surgeons with more accurate anatomical information.

Infective Endocarditis

Infective endocarditis is a rare but potentially life-threatening disease associated with intravenous drug use, prosthetic valve replacement, congenital heart disease, human immunodeficiency virus, and poor dentition. When involving the mitral valve, infectious vegetations typically form on the left atrial side of the mitral valve. Transeosophageal echocardiogram is the most reliable means of identifying lesions as mobile echogenic masses implanted in a valve, mural endocardium in the trajectory of a regurgitant jet, or in prosthetic material with no alternative anatomical explanation. On CT, mitral vegetations > 10 mm in diameter are almost always detected. They appear as a hypoattenuating, mobile mass adhering to the valve leaflet. On MRI, large vegetations typically appear as low signal masses attached to a valve leaflet and are almost always associated with mitral regurgitation. MRI is also useful to detect embolization of large vegetations.

Additional diagnostic considerations

While metastases represent the most common cardiac neoplasms, they were not included in this differential based on the imaging appearance of the mass in the illustrative case, as well as the clear association with the mitral annulus.

Diagnosis

Caseous calcification of the mitral annulus (CCMA)

Summary

The differential diagnosis for a mitral annular mass includes a variety of entities, to include CCMA, myxoma, peri-annular blood cyst, abscess, and infective endocarditis. The imaging appearance of the cardiac lesion presented in this case favors CCMA based on the lesion morphology, location, and presence of rim calcifications. CCMA may be distinguished from myxoma based on the fact that myxoma is usually mobile, pedunculated, and attached to the myocardial wall. Unlike CCMA, myxoma lacks peripheral calcifications, rarely involves the valve, and is highly vascular. Blood cysts appear as hypoechoic cystic lesions on ultrasound with smooth walls and hypodense cystic masses on CT with or without wall calcification; when wall calcifications are present, the lesions can mimic CCMA. Mitral annular abscess would not be expected to show extensive peripheral calcifications on CT. Lastly, in cases of infective endocarditis, vegetations typically affect the valve leaflet, rather than mitral annulus.

References

Abnormal Myocardial Enhancement

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Case Presentation

A 21-year-old man presented to the emergency department with fever and chest pain. Chest radiographs were normal. Electrocardiogram showed elevation of the ST-segments in leads II, III, and a VF, as well as reciprocal depression in leads I and a VL. Serum tests showed elevation of troponin T, creatine kinase (CK), and CK-MB. Echocardiography performed in the emergency department measured the left ventricular ejection fraction at 48%. Cardiac MRI with and without contrast was obtained to evaluate for myocarditis (Figure 1).

**FIGURE 1.** Delayed contrast-enhanced MRI image of the heart in short axis (A) demonstrates significant left ventricular epicardial hyperenhancement (arrow), particularly within the inferolateral walls. Three-chamber contrast-enhanced view of the heart (B) demonstrates additional foci of scattered midmyocardial hyperenhancement, particularly within the septum (arrow).
Key Imaging Finding(s)
Abnormal delayed epicardial and midmyocardial hyperenhancement

Differential Diagnoses
Infectious/inflammatory cardiomyopathy
Ischemic cardiomyopathy
Hypertrophic cardiomyopathy
Sarcoidosis
Amyloidosis

Discussion
Delayed contrast-enhanced MRI images of the heart demonstrate significant left ventricular epicardial hyperenhancement, particularly involving the inferolateral walls along with foci of scattered midmyocardial hyperenhancement, particularly involving the septum. Given the patient’s age, EKG changes and clinical symptoms of fever and chest pain, non-ischemic cardiomyopathy is the favored diagnosis. Examples of nonischemic cardiomyopathy include infectious cardiomyopathy, hypertrophic cardiomyopathy, sarcoidosis and amyloidosis. While ischemic cardiomyopathy is not favored, it is to be excluded whenever delayed contrast-enhanced cardiac MRI images are obtained.

Infectious Cardiomyopathy
Inflammation of the myocardium, or myocarditis, can result from several infectious, systemic or toxic etiologies. The majority of infectious processes are viral with the Coxsackie B virus responsible for about 50% of cases. The most common systemic processes to result in myocarditis include systemic lupus erythematosus (SLE), inflammatory bowel disease, and mixed connective tissue diseases. Toxic exposures from hydralazine, procainamide, heparin, warfarin, and anthracyclines are another source of myocarditis. The clinical picture of myocarditis is often vague; therefore, MR imaging may help establish a diagnosis or occasionally guide endomyocardial biopsy, when necessary. When there is a clear diagnostic picture with self-limited disease, however, cross-sectional imaging is usually not required. The typical MRI appearance of myocarditis is delayed ventricular wall enhancement that does not conform to a vascular distribution. Myocarditis typically affects the epicardial region of the myocardium with the lateral free wall most often involved.\footnote{1,3} Enhancement becomes more diffuse and faint over time, with or without associated wall motion abnormalities. As expected, there is often associated T2 hyperintense signal from inflammation. Pericardial effusions can occur but are often small.

In the chronic setting, fibrous tissue can replace inflammatory cells leading to an enhancing scar, similar to ischemic cardiomyopathy. The distinction from ischemic cardiomyopathy may be made if the scar does not conform to a vascular distribution or if there is preferential subepicardial involvement, as opposed to subendocardial involvement seen with ischemic disease. Involvement of the cardiac apex can be particularly difficult to distinguish from ischemic disease.

Ischemic Cardiomyopathy
Myocardial scarring and wall motion abnormality in a coronary artery distribution are the hallmarks of ischemic disease. The degree of wall involvement determines the likelihood of recovering contractility after reperfusion. Subendocardial infarction and scar involving < 50% of the wall thickness is associated with reversible ischemia of the remaining myocardium, which can regain or improve contractility when corrected (often with reperfusion procedures, such as stenting or revascularization). Transmural enhancement, on the other hand, is associated with decreased likelihood of recovery.\footnote{4} In chronic cases, transmural infarctions will typically show myocardial thinning, while subendocardial infarctions frequently have normal wall thickness.

Hypertrophic Cardiomyopathy
Hypertrophic cardiomyopathy (HCM) is the most common inherited cardiac disorder and is autosomal dominant in most cases. HCM is characterized by diffuse or focal left ventricular wall thickening, > 1.5 cm at end diastole but typically closer to 2.5 cm. Asymmetric interventricular septal involvement is the most common pattern; however, symmetric, apical (first described in Asian populations), mass-like, and end-stage or burned out (similar in appearance to dilated cardiomyopathy) forms also occur. Right ventricular involvement occurs in around 20% of patients. In addition to myocardial wall thickening, important MR imaging findings include midmyocardial patchy delayed enhancement – in particular at the right ventricular insertion points in the septum – in about 80% of patients, regional wall dyskinesia, and systolic anterior motion (SAM) of the mitral valve contributing to subaortic outflow tract obstruction.\footnote{3,5,6,7}

Sarcoidosis
Sarcoidosis is a multisystem non-necrotizing (noncaseating) granulomatous disease that most commonly involves the lungs and lymph nodes. It presents in young to middle-aged adults with a higher incidence in African-Americans and slight increased incidence in women. Approximately 5% of sarcoid patients will develop symptomatic cardiac disease, although up to 50% of patients have myocardial involvement on autopsy. Symptomatic patients often present with heart block, restrictive cardiomyopathy, and/or dysrhythmias and are at increased risk for sudden death. Delayed enhancement is typically patchy with involvement of the midmyocardium or subepicardium. The basal septum and lateral left ventricular free wall are often involved with sparing of the papillary muscles and right ventricular wall. Both chronic sarcoid and severe myocarditis can have transmural, whole-heart hyperenhancement, which may be distinguished from chronic ischemic disease by the lack of myocardial thinning. Sarcoidosis can also mimic myocarditis with subepicardial enhancement.\footnote{3,9}
**Amyloidosis**

The heart is affected pathologically in up to 90% of systemic light chain (AL) amyloidosis patients. Systemic amyloid AL amyloidosis is associated with monoclonal plasma cell dyscrasias, including multiple myeloma. Asymptomatic ECG abnormalities, followed by restrictive cardiomyopathy and congestive heart failure, are common clinical manifestations. The characteristic MR imaging appearance of amyloidosis consists of diffuse left ventricular subendocardial delayed enhancement with biventricular myocardial thickening. Diffuse biventricular delayed enhancement is another common appearance. A classic pitfall in amyloid imaging can occur during selection of inversion time to null the myocardial signal due to the intrinsic T1 prolongation caused by amyloid protein. With diffuse amyloid involvement of the myocardium, significant enhancement may be missed and mistakenly imaged as normal due to inappropriate nulling of this signal.

**Diagnosis**

Infectious cardiomyopathy

**Summary**

The pattern of delayed enhancement on cardiac MRI, when correlated with the appropriate clinical context and additional MRI findings, can typically differentiate nonischemic from ischemic cardiomyopathies. Delayed enhancement affecting the midmyocardial and epicardial regions in a nonvascular distribution is characteristic of nonischemic cardiomyopathies, while ischemic cardiomyopathies tend to have subendocardial or transmural delayed enhancement in a vascular distribution with associated wall motion abnormalities.

**References**

Hypertrophic Cardiomyopathy

A 60-year-old woman presented with chest pain. Echocardiogram was consistent with hypertrophic cardiomyopathy (HCM) and cardiac MRI was requested to better define the pattern of the disease.

EKG-gated cardiac MRI four-chamber views (fast imaging employing steady-state acquisition [FIESTA]) diastole [A] and systole [B] and late gadolinium enhancement [C] demonstrate marked midventricular hypertrophy (white arrowheads, A and B) with sparing of the apex (black arrowhead). There is marked left ventricular cavity narrowing (B). Confluent subendocardial to midmyocardial circumferential delayed enhancement is present in the midlateral and apical segments (arrows, C). At end diastole, the interventricular septum shows more severe thickening relative to the other segments and myocardial mass is markedly increased. Delayed apical enhancement suggests progression to the “burned out apex” with eventual apical aneurism formation.1

Differential diagnostic considerations include athlete’s heart, infiltrative cardiomyopathy, and aortic stenosis. The wall thickness in athlete’s hearts is less pronounced than that of HCM. Concentric wall thickening found with infiltrative cardiomyopathy and aortic stenosis differs from the predominantly septal thickening found with HCM. MRI can accurately identify the presence and distribution of myocardial thickening and define the morphological variant of HCM.2 The most common MR diagnostic feature is left ventricular wall thickness ≥ 15 mm at end diastole.1 The extent of myocardial fibrosis characterized by late gadolinium enhancement can serve as a predictor of major dysrhythmia.1

References
Left Ventricular Noncompaction

A 55-year-old man with a history of biliary colic necessitating stent placement presented to the emergency department with exertional chest pain. Physical exam was consistent with fluid overload. Preliminary laboratory data revealed minimally increased cardiac enzymes and an elevated creatinine.

The patient underwent a transthoracic echocardiogram (TTE), which demonstrated a markedly dilated left ventricle, severely reduced systolic function, and diffuse hypokinesis with an ejection fraction of 27%. At the apex of the apical four-chamber view in end systole (A), the ratio of the noncompacted left ventricular myocardium (between arrows) to compacted myocardium (between arrowheads) exceeds 2:1, consistent with echocardiographic diagnostic criteria for left ventricular noncompaction (LVNC). A four-chamber fast imaging employing steady-state acquisition (FIESTA) image through the apex in end diastole from an EKG-gated cardiac MRI (B) confirms the elevated ratio consistent with LVNC. No delayed myocardial enhancement was seen on post-contrast sequences.

A short-axis FIESTA image with a large field of view shows the findings of LVNC in a different plane (white double-headed arrow), as well as numerous left renal cysts (black arrows), pancreatic ductal dilatation (black arrowheads), and pneumobilia (white arrow heads) associated with patient’s known autosomal dominant polycystic kidney disease (ADPKD) and prior biliary stent placement.

LVNC is a congenital disorder of myocardial development with genetically heterogeneous defects associated with diverse protein-related gene mutations. Imaging criteria of LVNC are based on determining the ratio of the noncompacted to compacted myocardium, with the midlateral, apical, and midinferior segments most commonly involved. There is association between ADPKD and LVNC genetic disturbances.

References