

# Aberrant Right Subclavian Arteries

A review of the etiology, incidence, clinical presentation, and rationale for repair.

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**A**ortic arch abnormalities are an important factor to consider before endovascular surgery because solutions for aneurysms involving the distal and mid arch are now more widely available. These include new off-the-shelf branch options from a variety of endovascular platforms that can incorporate one branch into a repair without the need for customized manufacturing. Although the debate over the past decade has centered on the need to revascularize the left subclavian artery and the durability of mating branch stents in the proximal carotid artery, more common anatomic abnormalities can now also be considered for endovascular repair. This article reviews the literature on a common arch abnormality—the aberrant right subclavian artery and Kommerell diverticulum—and explores the indications for repair.

## DESCRIPTION, ORIGIN, AND PREVALENCE

The most common anatomic configuration of the aortic arch in humans is three supra-aortic trunks: the brachiocephalic, the left carotid, and the left subclavian. The aortic arch is embryologically formed when the persistent right proximal dorsal aorta fuses with the right seventh intrasegmental artery to become the right subclavian artery. When the arch is anomalous, an aberrant right subclavian artery occurs in 0.07% to 0.2% of patients with left-sided aortic arches.<sup>1-3</sup> Embryologically, the difference occurs when the fourth right aortic arch and distal aorta involute, and the seventh right intersegmental artery persists. The resulting subclavian artery is distinct from the right carotid and usually originates in the descending thoracic aorta distal to the left subclavian artery.

The aberrant course of the subclavian artery in this abnormality travels across the midline to the right arm. This has been termed the *lusorian artery* in some historical texts and was first described by Hunauld,<sup>4</sup> although Arkin is credited with coining this term.<sup>5</sup> This artery can pass behind the esophagus, between the trachea and esophagus, or in front of the trachea in

different presentations. Pediatric descriptions may also reveal a vascular ring. The symptomatic presentation of a right aberrant subclavian artery occurs when the artery exerts a mass effect on the trachea, esophagus, or other proximate structures, which was called *dysphasia lusoria* when it was first described in 1789 by Bayford.<sup>6</sup>

When the origin of the aberrant right subclavian artery is ectatic, it resembles the anatomic variation first described by Kommerell in 1936, now known as a Kommerell diverticulum.<sup>7</sup> Since that time, the term *Kommerell diverticulum* has been used to describe the aneurysmal origins of any aberrant subclavian artery (including an aberrant left subclavian artery originating in a right-sided aortic arch), as well as the diverticulum that results in nontraumatic aneurysms arising from the aortic isthmus. Other anatomic variants may accompany the aberrant right subclavian artery: truncus bicaorticus (19.2%), Kommerell diverticulum (14.9%), and aneurysm of the distal right subclavian artery (12.8%) and right-sided aortic arch (9.2%), but their incidence depends on the age of the population being studied.

## INCIDENCE AND PRESENTATION

A review of 1,378 cases of aberrant subclavian artery derived from the published literature on both clinical experience and autopsies in the pediatric age group was published in 1978 by Molz and Burri.<sup>8</sup> A female predominance (58%) was described, as well as a largely asymptomatic presentation, which are two features of this anomaly that continue to hold true. An isolated right aberrant subclavian artery without aneurysm in a left-sided aortic arch was found in 80% of cases. In this pediatric population, the presence of coarctation was far more prevalent than in later articles describing the same disease in adults.

A more modern review by Polguy et al in a predominantly adult population found 141 cases of aberrant right subclavian arteries in 796 abstracts.<sup>9</sup> The authors also described a female predominance (55.3% female vs 44.7% male), and the mean age of symptom onset was  $49.9 \pm 19.4$  years.<sup>9</sup> In adults, the aberrant subclavian

artery usually follows a path posterior to the esophagus (80%) but may also course between the esophagus and trachea (15%) or, rarely, anterior to the trachea (5%).<sup>10,11</sup> The most common presenting complaints were dysphagia (71.2%), dyspnea (18.7%), retrosternal pain (17%), cough (7.6%), and weight loss (5.9%).<sup>9</sup>

The most recent literature review by Tanaka et al<sup>1</sup> searched articles from 2004 to 2014, including a small number of autopsy studies for which the authors attribute the smaller aneurysm size. The age at presentation was  $41.8 \pm 26.6$  years. Fewer than half of patients with this anomaly will present with symptoms that can be attributed to the lusorian artery. Of these symptomatic patients, the most common presentations were dysphagia (34%), dyspnea (25%), chest pain (16%), abnormal chest x-ray (14%), back pain (6%), and arm claudication (5%). Ruptured diverticulum occurred in 4% of this cohort, and dissection of the aorta was found in 11%.<sup>1</sup> Kouchoukos et al and Idrees et al both reported a higher rate of dissection in the presence of this anomaly (20% and 50%, respectively).<sup>12,13</sup> A review of aberrant left subclavian arteries in the setting of a right-sided aortic arch was performed by Cina and colleagues, which provides a thorough description of this related abnormality.<sup>3</sup>

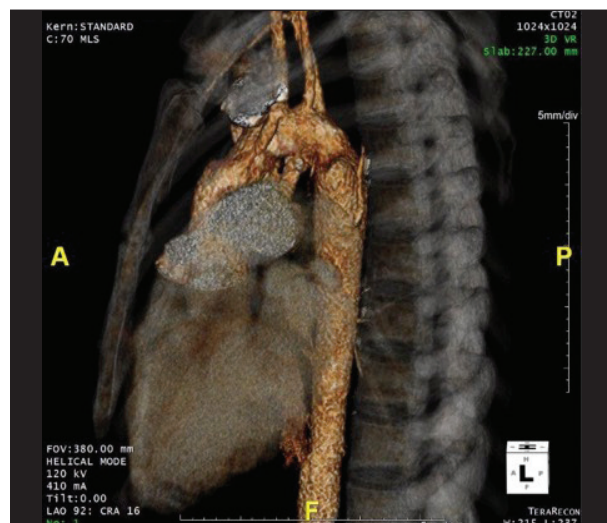
### MEASURING A KOMMERELL DIVERTICULUM

One of the largest barriers to determining the role of intervention in repair of these abnormalities is accurately describing the diverticular size. In initial descriptions of an ectatic origin of a right subclavian artery, Backer et al<sup>14</sup> used a measurement of 1.5 times the size of the distal subclavian artery as the criteria for determining the presence of an aneurysm. Although this may fit the criteria for a subclavian artery aneurysm, it belies the involvement of aortic tissue in the pathologic degeneration of this arterial segment. The findings of cystic medial necrosis in resected diverticula support the theory that a diverticulum is created with portions of attenuated aortic arterial wall intended to be involuted<sup>15</sup> and thus must be considered in the context of an aortic aneurysm as well. For this reason, the strategy proposed by Tanaka et al provides the most descriptive method of measurement, taking into consideration the diameter of the orifice of the diverticulum, as well as the distance from the furthest aortic wall to the end of the diverticulum.<sup>1</sup> Describing the diverticulum in two dimensions provides a better understanding of the pathology.

### INDICATIONS FOR REPAIR

#### Rupture and Aortic Emergencies

One of the most challenging issues to tease out of the heterogeneous literature on the aberrant subclavian artery



**Figure 1.** An aberrant right subclavian artery originating distal to the left carotid artery, with a stenotic and diminutive left subclavian artery originating distally.

is the indications for repair. Although no author has advocated the prophylactic repair of aberrant subclavian arteries in the absence of symptoms or rupture, a Kommerell diverticulum will accompany a right subclavian artery in 20% to 60% of cases.<sup>1,10,14</sup> In one center's imaging experience with both left- and right-sided arch abnormalities, 43% of patients with an aberrant right subclavian artery (left aortic arch) and 100% of the patients with an aberrant left subclavian artery (right aortic arch) had a Kommerell diverticulum.

The increasing incidence of dissection (usually type A dissection) in the presence of this anomaly has led some authors to recommend early intervention if an aneurysm is present. In the most modern review,<sup>1</sup> the diverticulum was measured at 15 to 80 mm in all patients, with the diverticulum ranging from 20 to 60 mm in patients presenting with rupture and 25 to 75 mm in those presenting with dissection. Cina et al recommended treatment when the diameter of the diverticulum was 3 cm at the level of the orifice in a low-risk patient.<sup>3</sup> After offering the sizing paradigm, Tanaka et al added that a diverticulum size > 50 mm between the wall adjacent to the trachea and the opposite aortic wall would also be a good indication for repair.<sup>1</sup> In all cases, because the anatomy of these anomalies is highly variable, the decision to intervene should also consider a center's experience.

#### Mass Effect and Dysphagia Lusoria

The symptoms associated with aberrant right subclavian arteries, including dysphagia, dyspnea, arm claudication, and pain, may also represent an indication

for surgery. In this scenario, repairs are tailored based on patient symptoms and the unique anatomy. It is essential to perform high-quality cross-sectional imaging and possibly angiography to determine the anatomic subtleties prior to intervention, as a number of concurrent anomalies can be present. Figure 1 demonstrates a right subclavian artery originating distal to the left carotid artery and crossing midline with symptomatic effect; however, subsequent careful inspection of the anatomy clarified a stenotic origin of the left subclavian artery distally and arising from a diverticulum.

## TYPES OF REPAIR

The indication for repair of an aberrant right subclavian artery has some bearing on the method chosen for repair. Patients with symptoms related to mass effect require removal or depressurization of the “space-occupying lesion.” Although a complete description of the surgical options for these patients is beyond the scope of this article, many authors agree that depressurizing the artery and partial resection can be sufficient to relieve symptoms. Ligation alone has fallen out of favor because of the risk of arm ischemia, so transposition or bypass frequently accompanies this approach.<sup>3,16</sup> Endovascular plugs have been used to decrease the pressure in the aberrant subclavian artery for patients who are too high risk for thoracotomy.<sup>17</sup>

Patients with asymptomatic aneurysmal disease originating from the right subclavian artery need prophylactic treatment due to their risk of rupture or aortic dissection. In this case, an approach involving both aortic and subclavian repair is necessary to decrease the risk of rupture and dissection. Perhaps owing to the rare nature of this abnormality, there is more literature describing the conventional surgical approach to repair through a thoracotomy,<sup>1,15,18</sup> and endovascular approaches are reviewed suspiciously because of the limited experience and durability.<sup>19</sup> However, in the modern era, endovascular options are increasingly described, most often including the combination of aortic stenting and subclavian revascularization.<sup>20-22</sup> This approach is preferred to placing an endovascular plug in the diverticulum alone because it does not rely on landing in diseased tissue to achieve seal. Endovascular treatment of an aberrant right subclavian artery with and without Kommerell diverticulum was reviewed by Vucemilo et al, and they describe a variety of endovascular options to manage multiple anatomic configurations, with a low (10%) perioperative mortality rate.<sup>22</sup>

For aneurysms that are very proximal and where there is no landing zone because of the proximity to the other supra-aortic trunks, a frozen elephant

trunk technique may be used with success. However, as endovascular branched options for arch anatomy become commercially available, the experience with total endovascular solutions to this challenging anatomic problem will likely be more widely reported.<sup>23</sup>

## CONCLUSION

Aberrant right subclavian arteries are rare, but when present, can be accompanied by a Kommerell diverticulum, which requires intervention at > 3 cm in orificial diameter and > 5 cm in depth. When patients present with symptoms, they can be treated with both endovascular and open strategies. ■

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