Taking a New Look at Kommerell: Recent Insights on Aortic Diverticula

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**ABSTRACT:** Kommerell’s aortic diverticula are infrequent but probably not as rare as previously estimated. They remain a conundrum of sorts because of lingering uncertainties as to their nature, the potential for catastrophic complications, and the indications for treatment. Abandoning the term diverticulum and using “Kommerell aneurysm” as a descriptor instead would be most helpful. The ability to size these lesions precisely and reproducibly is felt to be crucially important, and has been a historical barrier to appropriate management. The measurement techniques illustrated herein emerge as best and should become standard. Classification of Kommerell lesions into types 1 and 2 is practically useful and with important implications as to involved anatomy and treatment options. Surgical treatment is the historical gold standard, but the required operative approaches tend to be complex and potentially risky. Emerging hybrid surgical-endovascular strategies are appealing and may prove safer for a significant number of patients presenting with aortic diverticula of all types.

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**Key words:** aortic arch, diverticulum, aneurysm, Kommerell, arch anomalies

The finding of an aortic diverticulum at the origin of the aberrant right subclavian artery (aRSA) was reported in 1936 by German radiologist Burckhard Friedrich Kommerell in a case where the diverticulum seemed to give rise to such vessel.1,2 This was the first clinical – not postmortem – diagnosis of an aRSA, a vascular anomaly first noted in 1735 but not fully associated with the clinical syndrome of dysphagia caused by extrinsic compression of the esophagus until 1761 when Bayford provided a complete description.3 The descriptive Latin term *lusus naturae* (freak of nature) was used to denote the anomalous anatomy and the absence of an intrinsic esophageal lesion. Autenrieth called it *dysphagia lusoria* and, in 1926, Arkin proposed the term *arteria lusoria* as an appropriate label for the aRSA.4

At present, Kommerell’s diverticulum – or, diverticulum of Kommerell – is the term used universally to characterize the presence of the aneurysm-like funnel-shaped widening at the origin and proximal-most segment of an aberrant subclavian artery – whether right or left. It results from maldevelopment of the aorta with failure of regression with persistence of a remnant of the fourth primitive right or left dorsal arch in cases of, respectively, left-sided “normal” arch or right-sided anomalous arch.5 The rare “double aortic arch” is a
related anomaly and a source of some confusion as it can also cause dysphagia lusoria.6

**CLASSIFICATION**

Salomonowitz et al7 have proposed a simple but most useful classification of aortic diverticula that should be adopted by all:

**Type 1:** Diverticulum associated with left (so-called “normal”) aortic arch and aRSA (Figure 1);

**Type 2:** Diverticulum in right (anomalous) aortic arch with aberrant left subclavian artery (aLSA) (Figure 2); and

**Type 3:** Diverticulum arising from the isthmus (ductal zone) of the thoracic aorta, not associated with the subclavian artery. This can be best described as a non-Kommerell (or ductal) diverticulum (Figure 3).

Type 1 diverticula are generally conical in shape, while those associated with a right arch (type 2) are often larger and more rounded in configuration.

**INCIDENCE**

A left-sided aortic arch with aRSA is said to occur in 0.7% to 2.0% of the population, and more rare still is the right-sided arch with aLSA at 0.04% to 0.4%. A diverticulum of Kommerell has been reported to be...
present in 20% to 60% of individuals with an aberrant subclavian artery.\(^4\)

The true prevalence of these anomalies is now felt to be considerably higher than historical estimates in light of current experience where such lesions can be easily uncovered and beautifully delineated with computed tomography (CT) and magnetic resonance (MR) imaging. Gender differences appear to exist as well with female predominance noted with left aortic arch-aRSA, and male predominance with right arch-aLSA.\(^8\) The anatomical transmediastinal course of aberrant subclavian arteries varies, going behind the esophagus in 80% of instances, between the trachea and the esophagus in 15%, and in front of the trachea in 5% of cases.\(^9\) Clinically, respiratory symptoms are most common in the pediatric population because of their compression-prone soft trachea, whereas dysphagia and blood pressure mismatch in the upper limbs tend to predominate in the adult population. At times, Kommerell lesions attain very large size and cause severe compressive symptoms (Figure 4). However, only 5% of adults with aberrant subclavian arteries are symptomatic, being a purely imaging-based incidental diagnosis for the vast majority. Patients with right side

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**Figure 2.** Type 2 Kommerell. Note right and left common carotid arteries arising individually as the two most proximal branches, as well as the normal right subclavian artery and the aberrant left subclavian artery. RSA, right subclavian artery; RCCA, right common carotid artery; LCCA, left common carotid artery; aLSA, aberrant left subclavian artery.
arch may be more prone to rupture than those with a normal-configuration left arch.\textsuperscript{10}

**NATURAL HISTORY AND SIZING**

The natural history of aortic diverticula remains poorly defined, mainly because of the rarity of the condition. But 4 cm or larger could be considered a critical size threshold for rupture when published reports of such cases are taken into account,\textsuperscript{4} leading to the recommendation for elective treatment of diverticula measuring >3cm.\textsuperscript{11} The somewhat unpredictable behavior and perceived high rupture risk have led a few authors to suggest a policy of elective treatment for all aortic diverticula. Unsurprisingly, many more favor a selective approach. These challenges are made all the worse by the notorious difficulties regarding size measurements as the techniques used remain nonstandardized and vary widely in the literature. Fortunately, the superb anatomical definition afforded by today’s advanced CT and MR imaging represent an enormous improvement. The measurement techniques proposed by Idrees et al\textsuperscript{12} are reasonable and practically useful (\textbf{Figure 5}), as are

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\textbf{Figure 3.} Non-Kommerell ductal diverticulum on a patient with right-sided arch.

\textbf{Figure 4.} 7 cm Kommerell’s aneurysm causing severe compression of the esophagus (A), associated with aRSA and left side arch (B).
their suggested size thresholds for elective treatment in the adult population: 5 cm or larger for total diameter, and >3 cm at the base. Total diameter is likely the more important of the two. These measuring strategies can be applied to all cases, including non-Kommerell type 3 ductal diverticula.

For pediatric patients, Backer et al have proposed to designate as significant (and worthy of treatment) any diverticulum that is more than 1.5 times the diameter of the subclavian artery distal to it.13

TREATMENT

A variety of surgical approaches have been reported for Kommerell diverticula, including median sternotomy alone, right thoracotomy alone, left thoracotomy alone, bilateral thoracotomy, and median sternotomy plus thoracotomy.14 Various operative strategies have been used for resection of the diverticulum and arch repair, with lingering concerns regarding technical complexity and risks of complications – including mortality.

Figure 5. Imaging of a left arch and aberrant right subclavian artery (A) on which the recommended measurement technique was applied (B): total diameter from apex to opposite normal aortic wall (a), diameter at the base where diverticulum originates off the aorta (b). Modified from Tanaka et al.4
HYBRID TREATMENT

Less invasive hybrid approaches that combine surgical and endovascular techniques have emerged in recent years. Management of “patient G” (Figures 6, 7) illustrates one such strategy. The patient was referred for treatment of a large type 2 Kommerell associated with aLSA. The author has treated 3 additional type 2-patients with the same general approach, but with an aorto-right axillary artery bypass (instead of carotid-axillary) being performed at the time of arch debranching.

Two additional hybrid strategies for aLSA and right arch are worthy of note. Planned antegrade delivery of the thoracic endograft to facilitate precise deployment across the sharply angulated anatomy is at the center of both:

- Single-stage frozen elephant trunk with antegrade delivery of the endograft through an open aortotomy approach\(^2\) (Figure 8); or
- Single-stage median-sternotomy with arch debranching and simultaneous antegrade delivery and deployment of the thoracic stent-graft across the arch to cover the aLSA (Figure 9).

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**Figure 6.** A 58-year-old male presenting with right side arch (and right descending thoracic aorta) and a large type 2 Kommerell lesion associated with aLSA. RSA, right subclavian artery; RCCA, right common carotid artery; LCCA, left common carotid artery; aLSA, aberrant left subclavian artery.
Vascular-plug closure of the subclavian artery just beyond the diverticulum is an important component for both.

Treatment of type 1 diverticula associated with aRSA and normal-configuration left-side arch tends to be more straightforward. The likely anatomic proximity of the aRSA to the origin of the normal LSA makes endograft coverage of both vessels frequently neces-

Figure 7. Staged repair consisted of, first, arch debranching via median sternotomy and side-graft bypass from the root of the ascending aorta to the right and left common carotid arteries. The second stage (several weeks later) involved a right carotid-axillary artery bypass, transfemoral endograft deployment across the arch, and left transbrachial vascular-plug closure of the LSA just beyond the diverticulum (A, B).
necessary, implying the need for preliminary debranching and revascularization of one or both. Closure of the aRSA beyond the diverticulum is required to secure complete exclusion (Figure 10). Such treatment strategy has been used successfully in three type-1 patients managed by the author.

Lastly, it is important to note the recently described subclavian periscope technique that can be used in the treatment of type 1 lesions, especially in situations where debranching and cervical bypass operations are deemed impractical or unfeasible (Figure 11).

**Figure 8.** Single-stage frozen elephant-trunk technique via open aortotomy in type 2 anatomy: median sternotomy and cardio-pulmonary bypass required. Note antegrade delivery of endograft (arrow) under direct visualization, and vascular-plug closure of left subclavian artery just beyond the Kommerell aneurysm. Modified from Idrees et al.12

**Figure 9.** Arch debranching and simultaneous antegrade delivery and deployment of thoracic stent-graft for type-2 lesions.

**Figure 10.** Endografting of arch and proximal descending thoracic aorta with coverage of the normal LSA and aRSA, and vascular-plug closure just beyond the aneurysm. Not illustrated is the required revascularization of one or both subclavian.
CONCLUSION

Aberrant subclavian arteries and Kommerell diverticula are relatively rare vascular anomalies, but their prevalence may well be considerably higher than previously suggested. The potential for rupture (and aortic dissection) is real and somewhat unpredictable, and these risks grow exponentially with increasing size. Adopting the term Kommerell’s aneurysm – instead of the more obscure “diverticulum” – would be helpful in the author’s view and likely lead to a better understanding of the clinical implications and the need for treatment. The measurement techniques herein illustrated are felt to represent the best approach to sizing and to clinical decision-making regarding treatment.

It is important and practically useful to classify Kommerell aneurysms into two main groups: Type 1 – normal-configuration left arch with aRSA; and Type 2 – anomalous right arch with aLSA. Even more rare and belonging with the latter from a repair-strategy viewpoint is the ductal non-Kommerell diverticulum (type 3).

Various surgical approaches have been reported for adult Kommerell patients, with much success in many cases. However, the involved operations are formidable and can only be performed with the required expertise and safety at very few centers of excellence around the world. The still-evolving hybrid strategies emerged in the effort to lessen invasiveness and lower risks. Several options are available, and reported results are so far encouraging. They can be portrayed as undoubtedly appealing, but careful analysis of a larger collective experience with longer patient follow-up must be awaited before we can declare with confidence that a new standard of care has arrived.

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