Re-visiting Kommerell:

Recent insights on management of *aortic diverticula*

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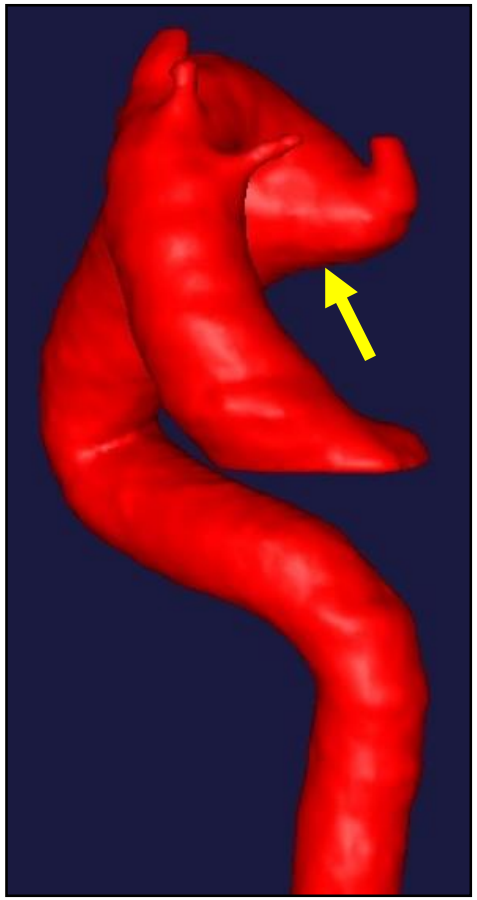
Disclosure of potential conflict of interest:

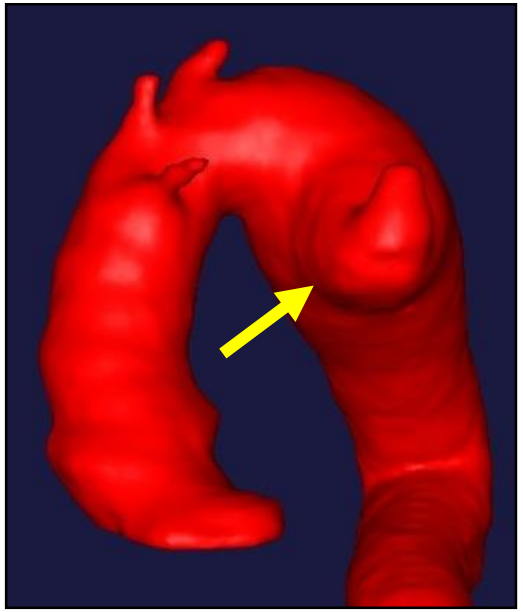
Frank J Criado

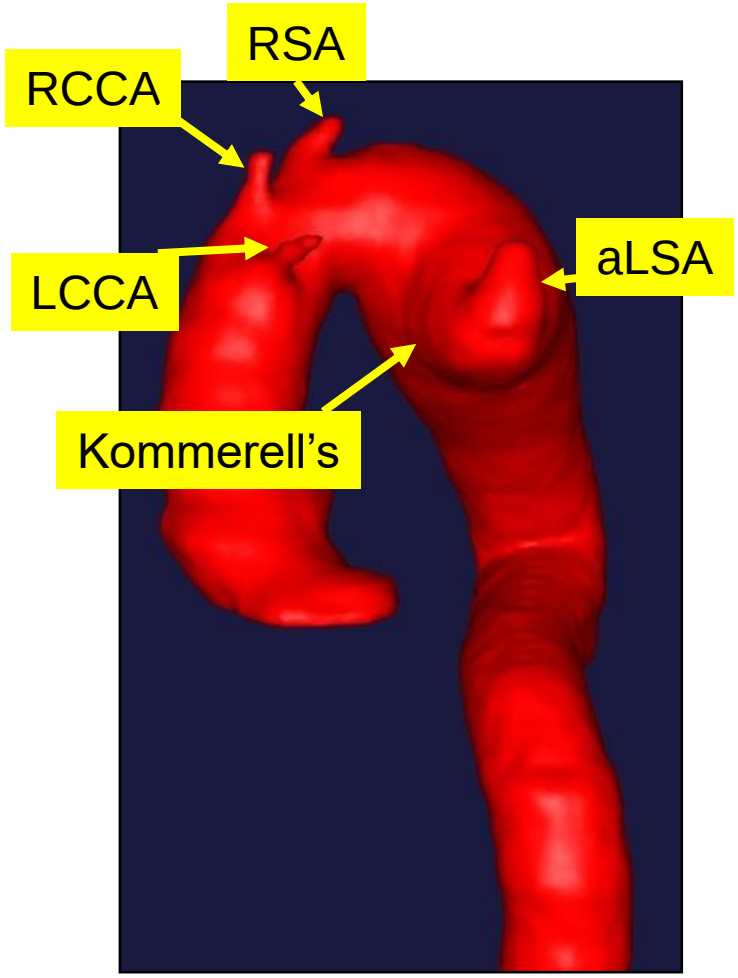
- **Medtronic:** honoraria for sales training, consulting

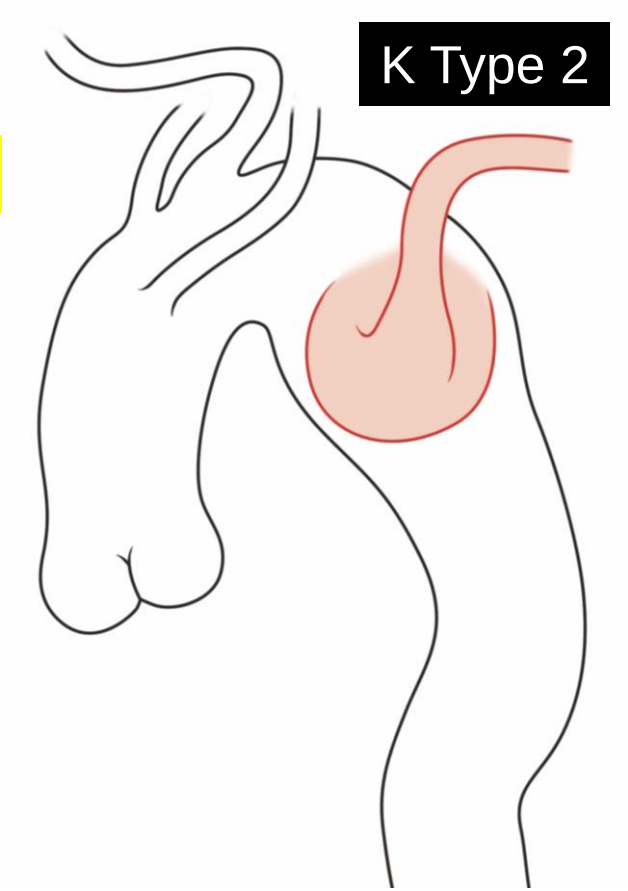


58y man referred for Rx of a large Kommerell's diverticulum associated with an aberrant LSA (aLSA) and right side arch (right side descending)

History of prior right postero-lateral thoracotomy and aborted attempt at surgical exposure and repair

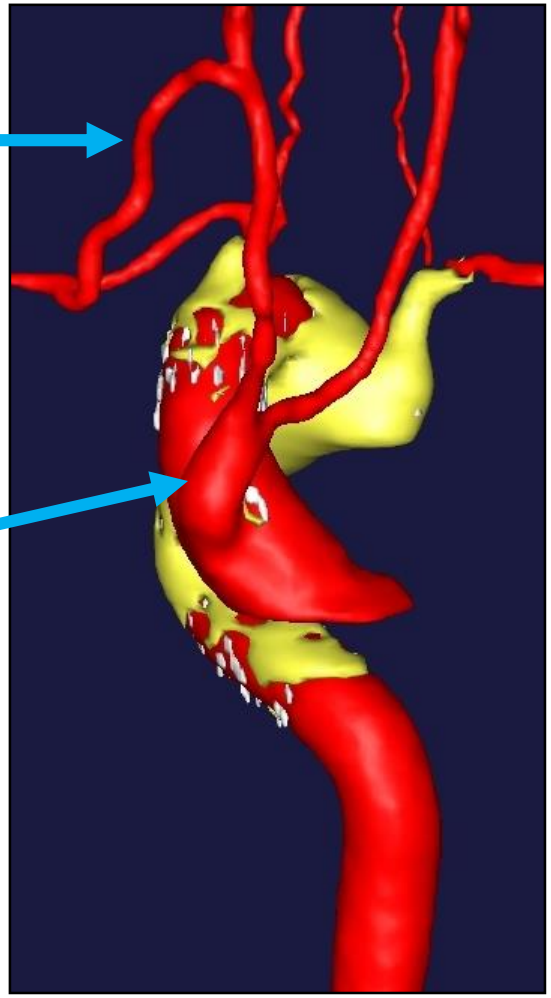


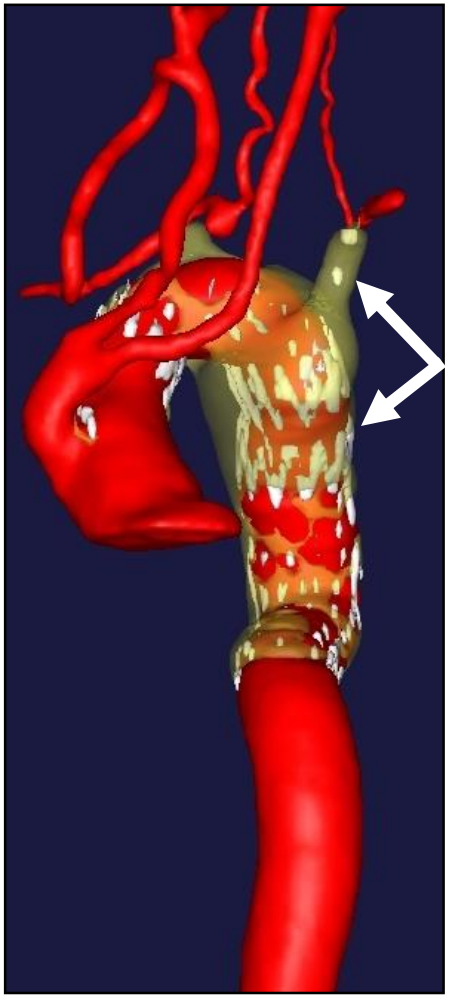

RSA
RCCA
aLSA
LCCA
Kommerell's


K Type 2

Staged hybrid repair:

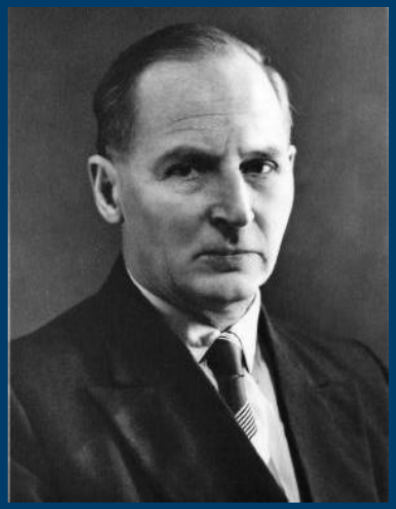
- Median sternotomy, asc aorta-based bypass to Rt+Lt CCAs
- Rt carotid-axillary bypass, trans-femoral endografting, vascular plug closure of aLSA



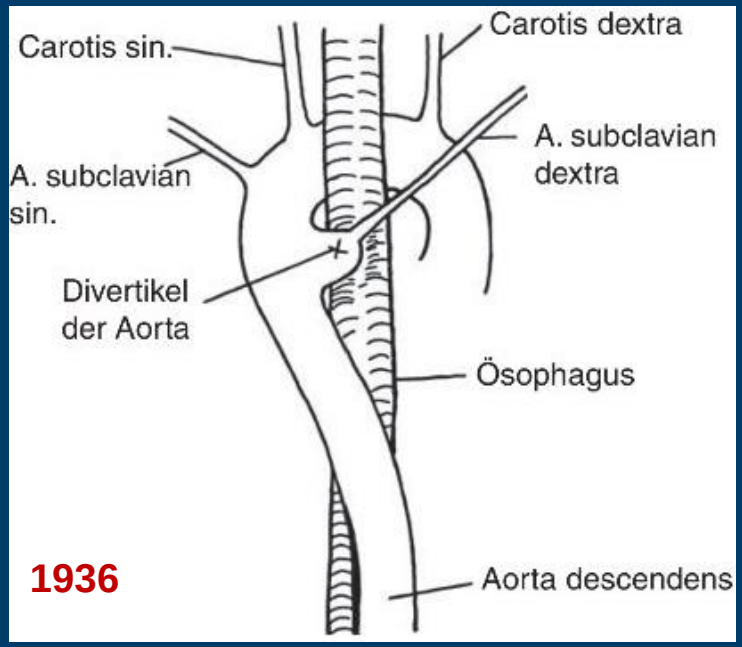


Trans-femoral endografting, vascular plug closure of aLSA

Kommerell Diverticulum



Burckhard F. Kommerell
1901-1990



1936

"Kommerell's diverticulum" – "Diverticulum of Kommerell"

1735 – first description of aRSA (Hunauld, at autopsy)
1761 – extrinsic compression of esophagus (Bayford)

Lusus naturae (freak of nature)

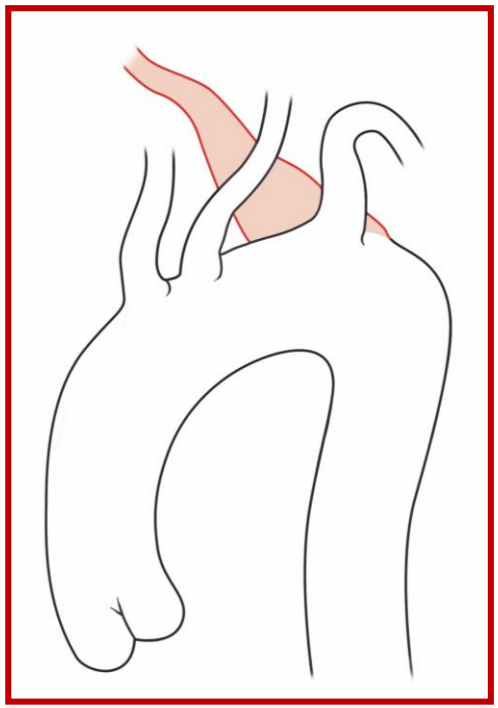
Dysphagia lusoria (Autenrieth, early 1800s)
Arteria lusoria (Arkin, 1926)

1936 – *Kommerell made first clinical diagnosis of esophageal compression by a posterior coursing lusorian artery with a diverticulum*

Classification and Incidence

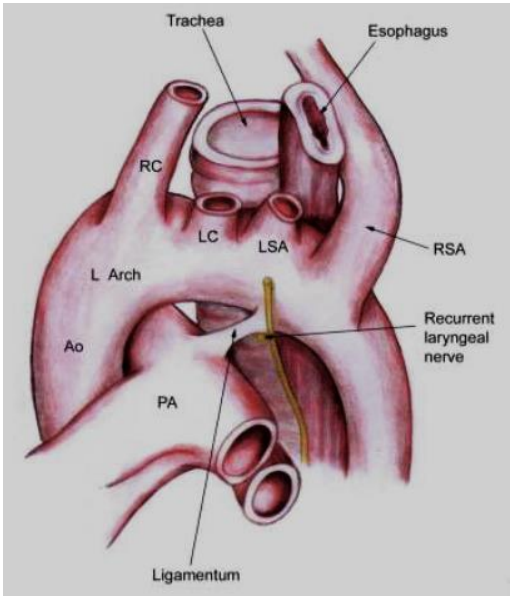
The classification of aortic diverticula proposed by Salomonowitz *et al*[7] is most useful and deserving of universal embrace:

- Type 1: Diverticulum occurring in left (so-called "normal") aortic arch in association with an aberrant right subclavian artery (Figure 1)



Reported <u>prevalence</u> of "normal-anatomy" left side arch with aRSA:
0.7-2.0% of the population

K Type 1



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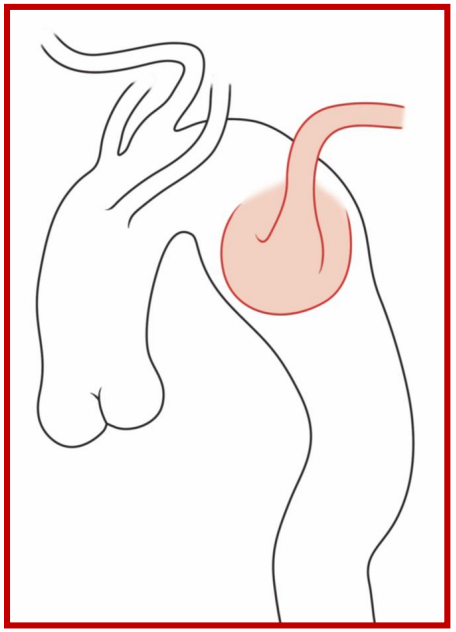
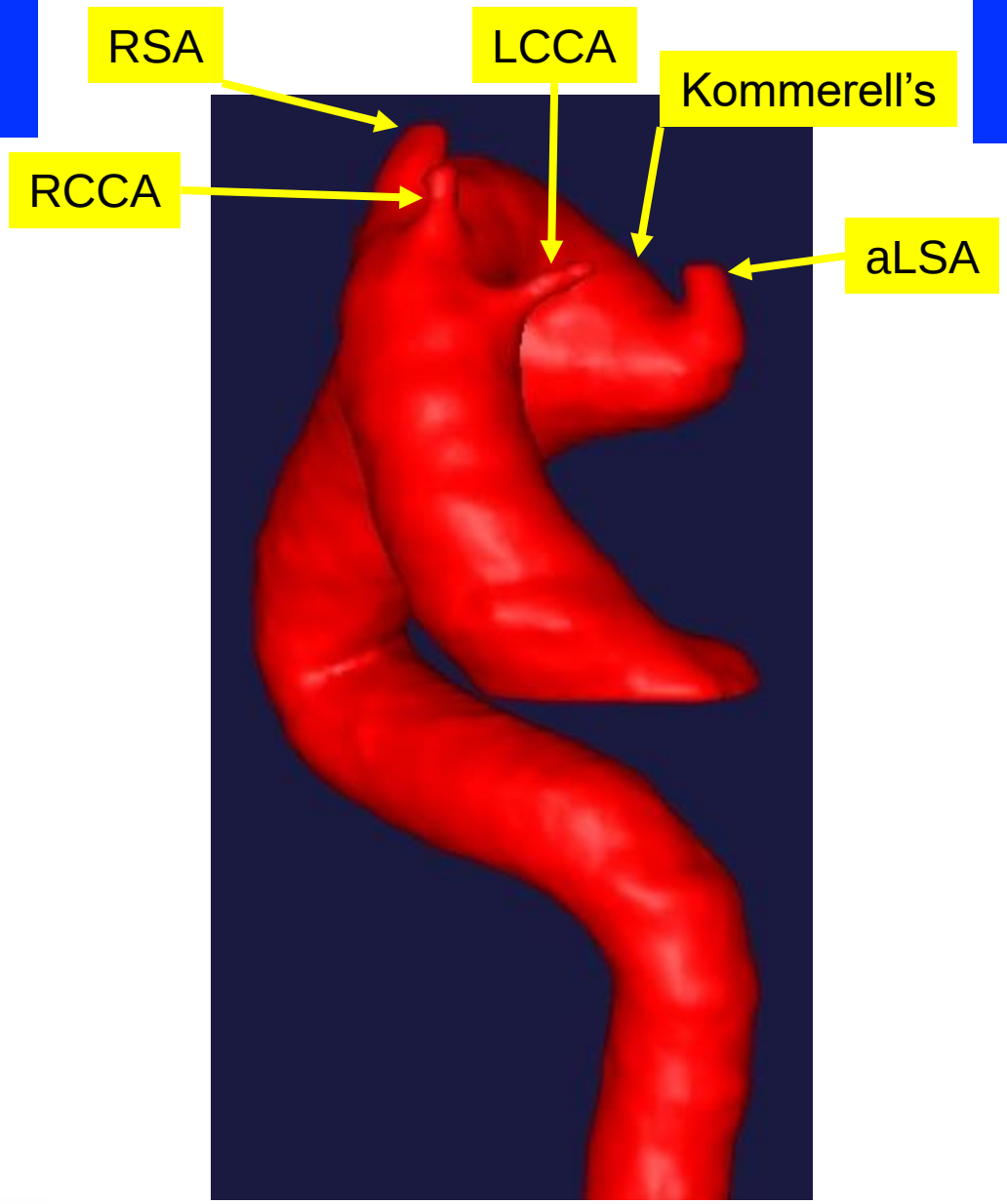
- Type 2: Diverticulum in right (anomalous) aortic arch associated with aberrant left subclavian artery (Figure 2)



Reported prevalence of right side arch with aLSA:
0.04-0.4% of the population

K Type 2




RSA
LCCA
Kommerell's
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aLSA

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- Type 2: Diverticulum in right (anomalous) aortic arch associated with aberrant left subclavian artery (Figure 2)
- Type 3: Diverticulum emerging from the isthmus (ductal zone) of the thoracic aorta, not associated with the subclavian artery: non-Kommerell (or ductal) diverticulum (Figure 3).

Type 3

Ductal or *Non-Kommerell*

The presence of a Kommerell's diverticulum has been reported in 20–60% of individuals with an aberrant subclavian artery.[4]

In terms of gender differences, female predominance has been noted with left aortic arch-aberrant right subclavian artery, and male predominance with right arch-aberrant left side artery.[8] Anatomically, aberrant subclavian arteries course behind the oesophagus in 80% of instances, and between the trachea and the oesophagus in 15%. A pre-tracheal course has been noted in 5% of cases.[9]

Aberrant subclavian arteries course:

- **behind the esophagus = 80%**
- between the trachea and esophagus = 15%
- in front of the trachea = 5%

Aberrant Subclavian Artery Pathologies and Kommerell's Diverticulum: A Review and Analysis of Published Endovascular/Hybrid Treatment Options

Chenzi Yang, MD[1]; Chang Shu, MD, PhD[1]; Ming Li, MD[1]; Quanming Li, MD[1]; and Reinhard Kopp, MD *J Endovasc Ther. 2012;19:373–382*

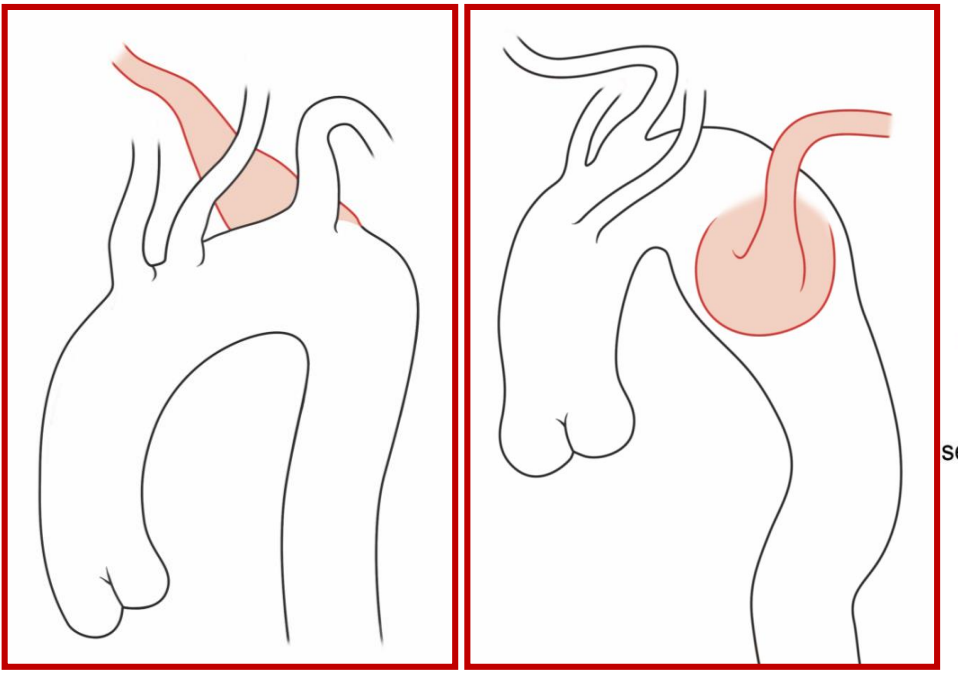
Gen Thorac Cardiovasc Surg (2015) 63:245–259
DOI 10.1007/s11748-015-0521-3

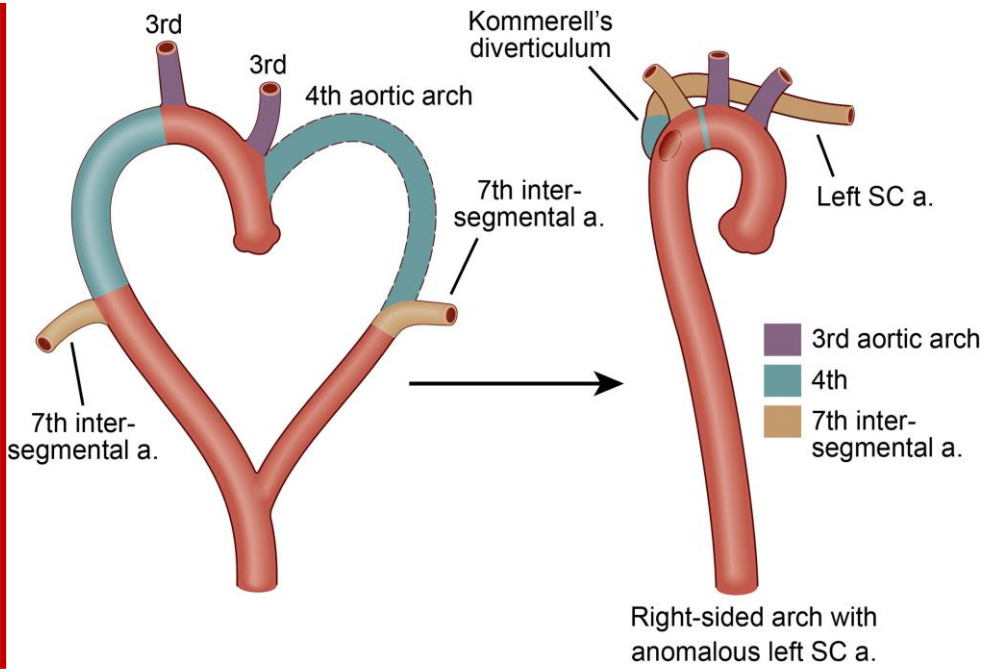
CURRENT TOPICS REVIEW ARTICLE

Kommerell's diverticulum in the current era: a comprehensive review

Akiko Tanaka · Ross Milner · Takeyoshi Ota

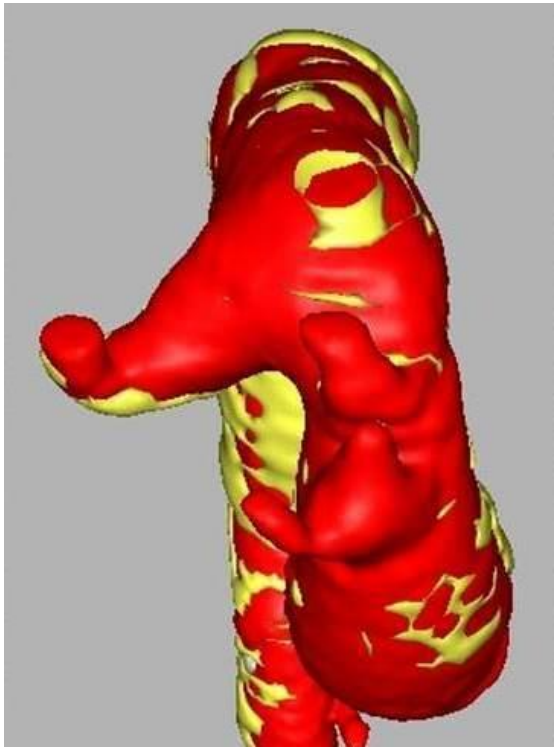
Kommerel's diverticulum is a remnant of the dorsal aorta from embryonic development, and **it is always associated with an aberrant SA**

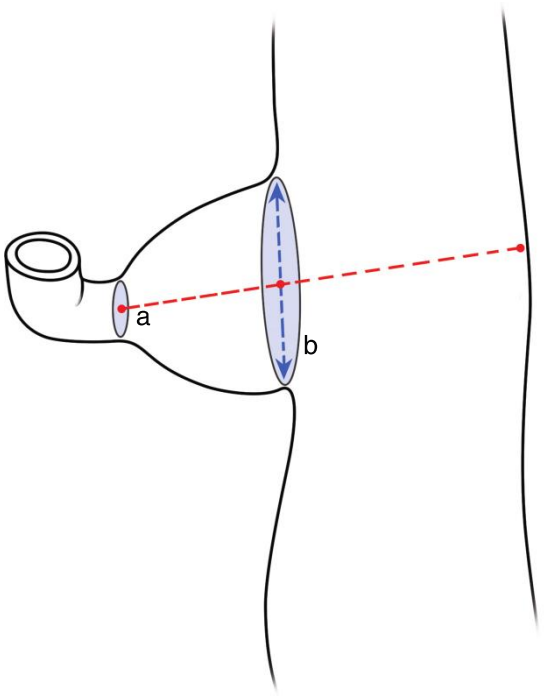




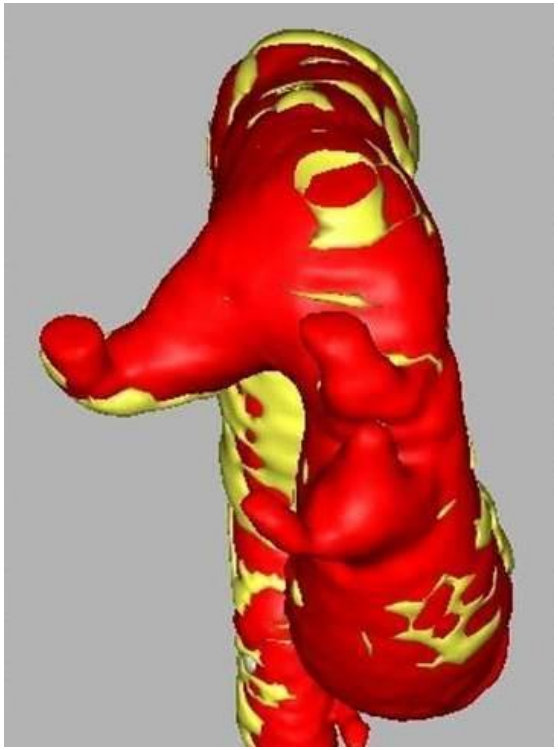
Natural history and sizing

The natural history of Kommerell's diverticulum remains undefined, mainly because of the rarity of the condition. The literature fails to provide clear-cut information on a critical size threshold for rupture, but 4cm or larger is the figure described in a preponderance of reported cases,[4] leading to the recommendation for elective treatment of diverticula measuring >3cm.[11] Difficult to justify as it may be, some authors espouse a policy of elective treatment for all aortic diverticula based on the unpredictable behaviour and perceived high rupture risk, but many more (this author included) favour a selective approach. Added to these challenges are the notorious difficulties regarding best sizing and measurement techniques as they remain non-standardised and vary widely in published reports. The situation is

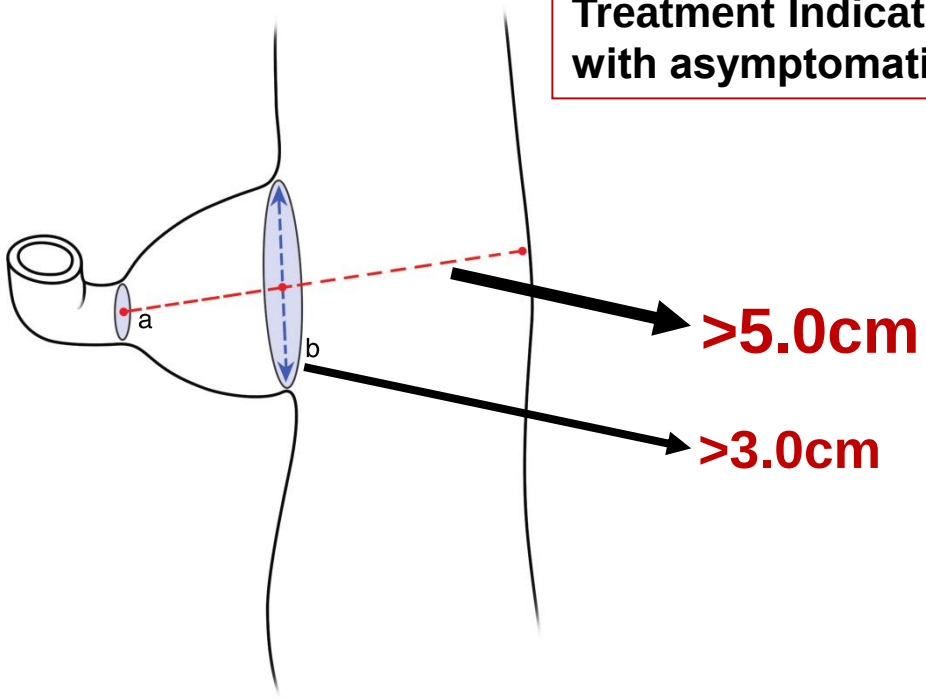


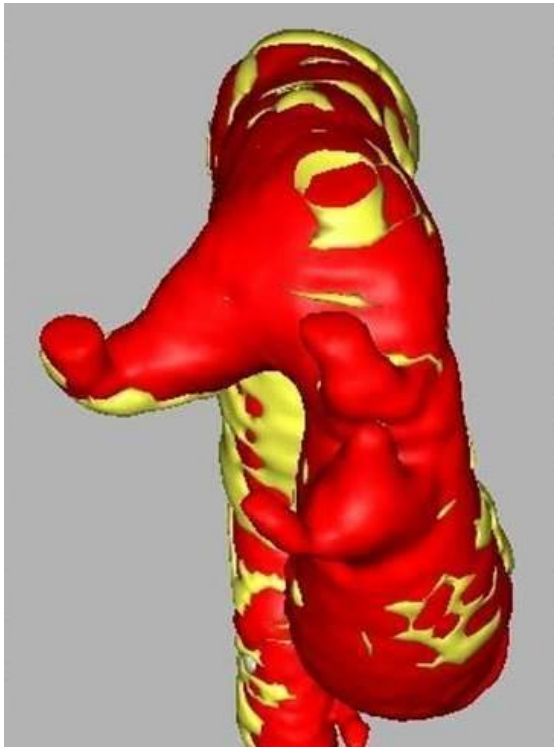


Measuring Kommerell

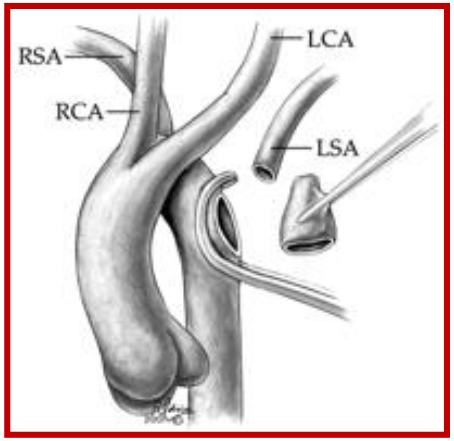


Treatment Indications for adults with asymptomatic Kommerell's





Kommerell patients presenting with <u>mediastinal compression symptoms</u>



Backer CL et al.
Eur J Cardiothorac Surg 2002

Table 1 Open surgical procedures

Procedure	Approach	*N* (emergent)	Rupture/ dissection	Lt/Rt arch	Reconstructions of the ASA	Additional cervical incision	Operative death	Complications
Graft replacement of the descending aorta[a]	Rt thoracotomy	11 (2)	1/2	1/10	In situ 6, bypass 3, translocation 1, none 1	4	1 (APTE)	Respiratory failure 1, APTE 1
	Lt thoracotomy	12 (1)	1/1	11/1	7 In situ, 4 bypass	4	1[b] (MOF)	Hoarseness 1, chylothorax 1 bleeding 1
Resection/patch closure/ aneurysmorrhaphy of the KD[c]	Midsternotomy	2 (0)	0/0	2/0	Bypass 2	0	0	Respiratory failure 1
	Rt thoracotomy	16 (0)	0/0	1/15	Translocation 9, bypass 4, in situ 1, none 2	13	0	Graft thrombosis 1, dissection 1, phrenic nerve injury 1, respiratory failure 1, TND
	Lt thoracotomy	50 (1)	0/0	6/44	Translocation 40, bypass 7, in situ 1	7	0	Hoarseness 2, phrenic nerve injury 1, chylothorax 4, bleeding 2, ptosis 1
Division of the ligamentum arteriosum[d] (untouched KD)	Midsternotomy	3 (0)	0/0	1/2	N/A 3	0	0	
	Lt thoracotomy	16 (0)	0/0	0/16	N/A 8, none 8	0	0	Persistent symptom 6, chylothorax 1
Total arch replacement	Midsternotomy	7 (3)	0/3	1/6	In situ 3, bypass 3, translocation 1	0	1 (Mediastinitis)	TND 1, mediastinitis 1
	Midsternotomy + Rt thoracotomy[e]	10 (0)	0/1	0/10	In situ 10	3	0	Mediastinitis 1, respiratory failure 1
	Bilateral thoracotomy[f]	4 (0)	0/0	2/2	In situ 3	0	0	Hoarseness 1

Table 2 Hybrid TEVAR and Total TEVAR

Procedure	Approach	*N* (emergent)	Rupture/ dissection	Lt/Rt arch	Procedures to the ASA	Operative death	Complications
Division of the ligamentum arteriosum + frozen elephant trunk (ASA not ligated)	Upper-J sternotomy + femoral	3 (1)	0/1	0/3	None 3	0	
Total arch replacement + elephant trunk + TEVAR	Midsternotomy + femoral	5 (0)	0/4	5/0	In situ reconstruction 4, coiling 1	0	
KD coiling/plug	Cervical incision + femoral	2 (0)	0/0	1/1	Translocation 1, bypass 1	0	
Total supra-aortic debranching TEVAR (zone 0)	Midsternotomy[a] + femoral	3 (0)	0/0	1/2	Coiling 1, coiling +bypass 1, none 1	1 (respiratory)	Respiratory failure 1
	Midsternotomy + cervical + femoral	4 (2)	1/0	1/3	Bypass 4	0	
Partial supra-aortic debranching TEVAR (zone 1)	Cervical incision + femoral	8 (1)	0/1	7/1	Coiling + bypass 5, bypass 2, coiling 1 none 1	0	Stroke 1
Partial supra-aortic debranching TEVAR (zone 2)	Cervical incision + femoral	10 (3)	2/1	8/2	Ligation + bypass 6, coiling + bypass 2, bypass 1, translocation 1	1[b]	Arterial-esophageal fistula 1, hoarseness 1
	Midsternotomy + cervical + femoral	1 (0)	0/0	0/1	Coiling + bypass 1[c]	0	
Total TEVAR (zone 2)	Femoral	6 (1)	0/1	2/4	Coiling 5, none 1	0	Arm claudication 1, arm pressure drop (50 mmHg) 1, type 2 endoleak 1
Total TEVAR with fenestrated endoprosthesis (zone 1–2)	Femoral	2 (0)	0/0	1/1	Reconstruction with branching 2[d]	0	

Hybrid repair of Kommerell diverticulum

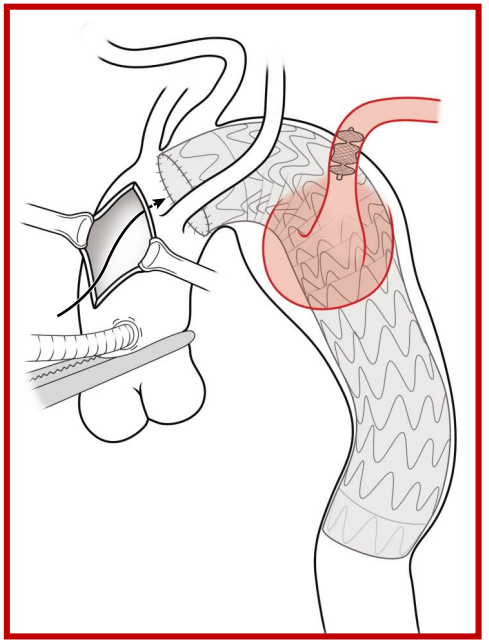
Jahanzaib Idrees, MD,[a] Suresh Keshavamurthy, MD,[a] Sreekumar Subramanian, MD,[a] Daniel G. Clair, MD,[b] Lars G. Svensson, MD, PhD,[a] and Eric E. Roselli, MD[a]

Conclusions: Hybrid repair is a safe and effective surgical treatment option for Kommerell diverticulum. Selection of the specific type of intervention is based on patient anatomy and comorbid conditions. (J Thorac Cardiovasc Surg 2014;147:973-6)

Hybrid strategies

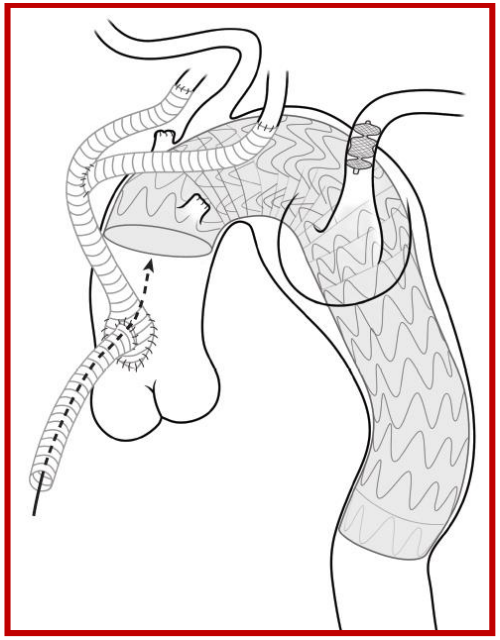
Two other hybrid strategies for diverticula associated with aberrant left subclavian artery in right arch cases are worthy of description. They both emphasise antegrade delivery of the thoracic endograft to facilitate precise deployment across the sharply angulated anatomy that often creates severe challenges when using the standard trans-femoral access technique:

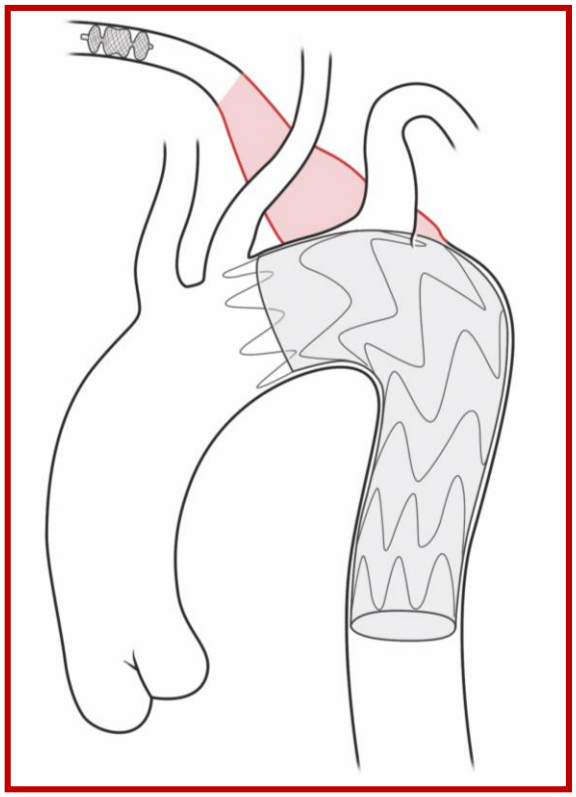
- Single-stage frozen elephant trunk with antegrade delivery of the endograft through an open aortotomy approach[12] (Figure 8) or

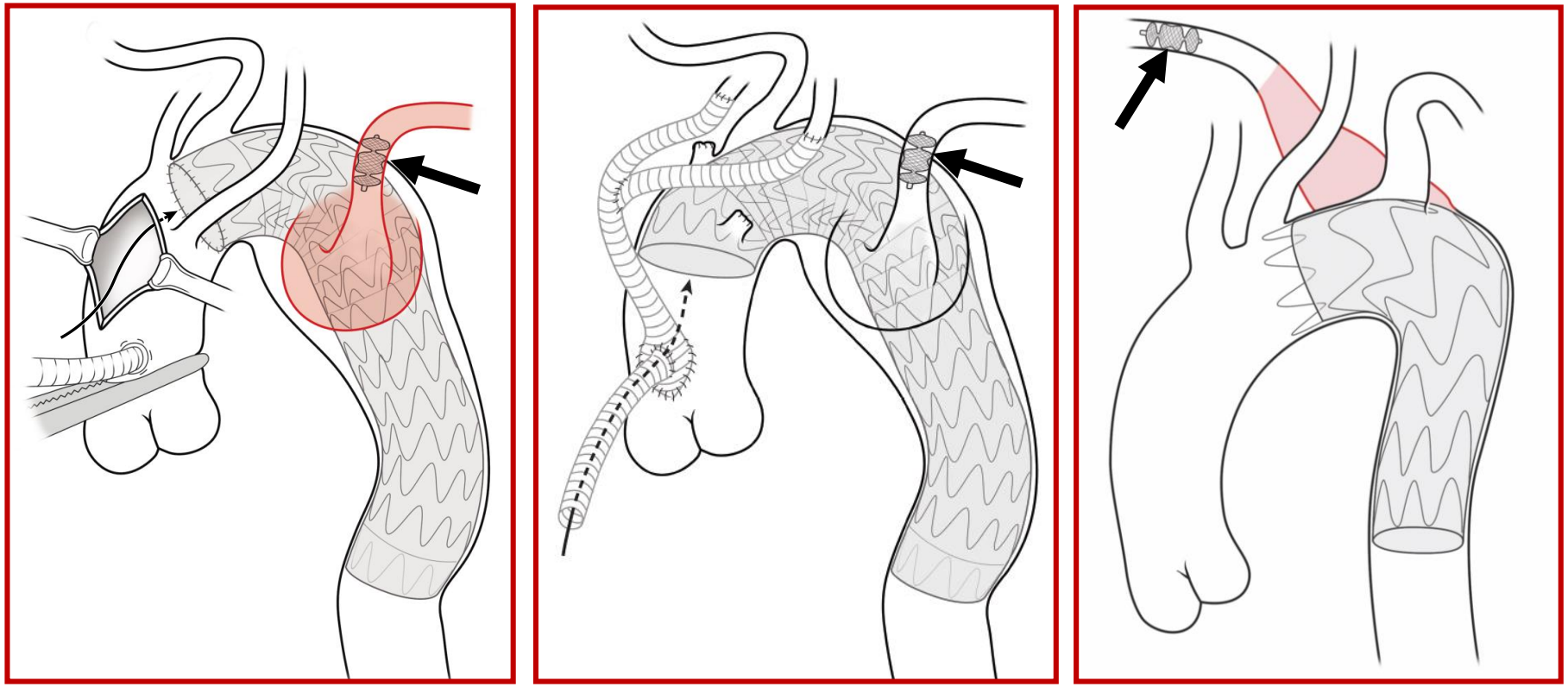


Modified from
Idrees et al. 2014

- Single-stage frozen elephant trunk with antegrade delivery of the endograft through an open aortotomy approach[12] (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 aberrant left subclavian artery (Figure 9).



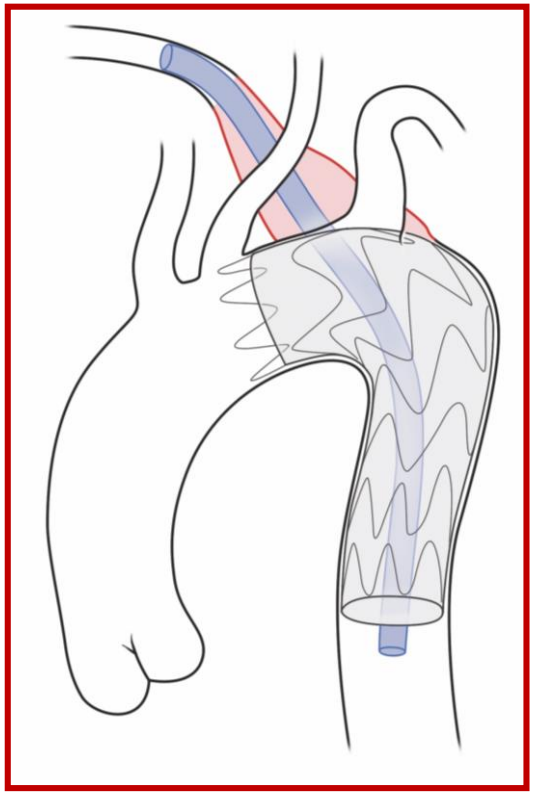




Closure of the involved SA just beyond the diverticulum (Vascular Plug or Coils) is an important common component to all these techniques

Trans-femoral aRSA

PERISCOPE



◆CLINICAL INVESTIGATION ◆

Periscope Endograft Technique to Revascularize the Left Subclavian Artery During Thoracic Endovascular Aortic Repair

Mario Lachat, MD[1]; Dieter Mayer, MD[1]; Thomas Pfammatter, MD[2]; Frank J. Criado, MD[3]; Zoran Rancic, MD, PhD[1]; Thomas Larzon, MD, PhD[4]; Frank J. Veith, MD[1,5]; and Felice Pecoraro, MD[1,6]

◆CASE REPORT ◆

Chimney and Periscope Grafts to Facilitate Endovascular Treatment of Aortic Transection in a Patient With Aberrant Right Subclavian Artery

Sandra Vicente, MD[1,2]; Michael Glenck, MD[3]; Dieter Mayer, MD[1]; Frank J. Veith, MD[4]; Mario Lachat, MD[1]; and Felice Pecoraro, MD[5]

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

Frank J. Criado, MD
From MedStar Union Memorial Hospital, Baltimore, Maryland.

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.

VASCULAR DISEASE MANAGEMENT 2016;13(7):E156-E165
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 post-mortem – 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

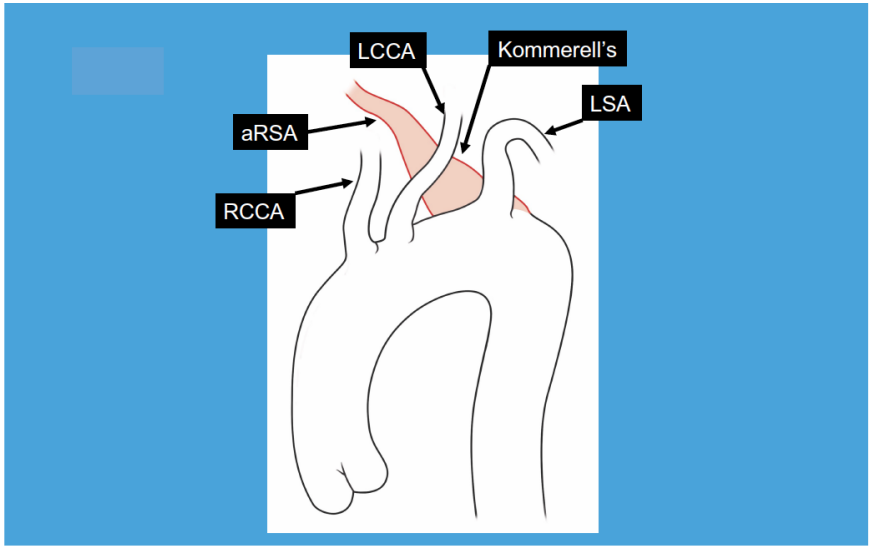


Figure 1. Type 1 Kommerell associated with a normal-configuration left arch and aRSA. aRSA, aberrant right subclavian artery; RCCA, right common carotid artery; LCCA, left common carotid artery; LSA, left subclavian artery.

related anomaly and a source of some confusion as it can also cause dysphagia lusoria.[6]

CLASSIFICATION

Salomonowitz et al[7] 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

Summary

- Kommerell's diverticula are infrequent but probably not as rare as predicted by previous estimates.
- They remain a conundrum of sorts because of lingering uncertainties as to their nature, the potential for catastrophic complications, and the indications for treatment.

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- Kommerell's diverticula are infrequent but probably not as rare as predicted by previous estimates.
- 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, instead, the proposed Kommerell aneurysm as a descriptor would be helpful.
- The ability to measure these lesions precisely and reproducibly is felt to be crucially important, and a historical barrier to appropriate management. The techniques described in the chapter emerge as best and should become standard.
- Classification of Kommerell lesions into types I and II is very helpful and with important implications as to anatomy and repair options.
- Surgical treatment is the historical gold standard, but the involved operative approaches are frequently complex and risky.
- Emerging hybrid surgical-endovascular strategies are appealing and may prove safer for some patients presenting with aortic diverticula of all types.