Hybrid Surgical Treatment of Dysphagia Lusoria in a patient with Loeys-Dietz Syndrome

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INTRODUCTION

- **Dysphagia Iusoria (DL)** results from the compression of the esophagus by an aberrant right subclavian artery (ARSA).
- Its prevalence is reported to be less than 1%.
- Management ranges from dietary modifications to surgical intervention in severe cases. Surgery entails transposition of the ARSA to the right common carotid artery (RCCA), and ligation and resection of the ARSA.





OBJECTIVE

The aim of this presentation is to describe an unusual case involving an aberrant right subclavian artery (ARSA) in a patient with Loeys-Dietz syndrome (LDS).





METHODS

- A 47-year-old woman was transferred to our institution with a 30-year history of **dysphagia** secondary to an ARSA.
- **Past medical history** was significant for LDS, Hashimoto's disease, and a craniotomy for the resection of a sphenoid meningioma.

• Pre-operative computed tomographic angiography (**CTA**) scan demonstrated an ARSA and a Kommerell diverticulum (figure 2).



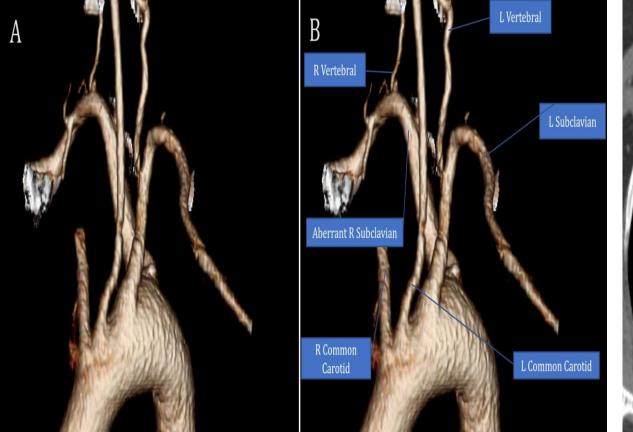


Figure 1. Fig 1A: ARSA. Retroesophageal course. Fig 1B: Supraaortic vessels.

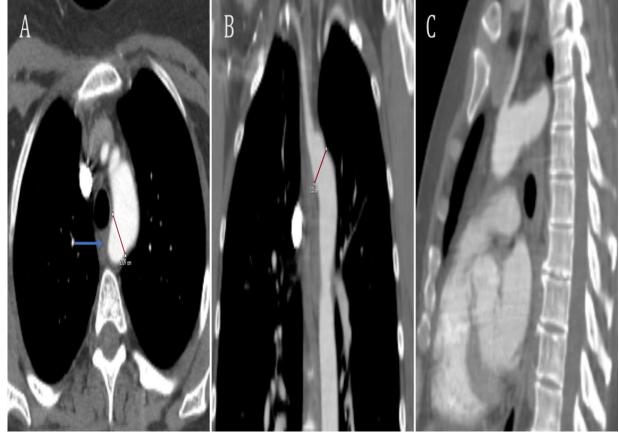


Figure 2. 2-cm Kommerell diverticulum at the site of the ARSA.



- **The 1**st **stage** was an ARSA transposition to the Right common carotid (RCCA) via a supra-clavicular approach.
- The ARSA was transected, and oversewn in 2 layers.
- Immediately after clamping the RCCA: artery dissection: Right carotid interposition graft + Right carotid- subclavian bypass performed.
- The RCCA was stented using a 14 x 60 mm self-expandable stent proximally and a 7 x 30 mm self-expandable stent distally (figure 3).



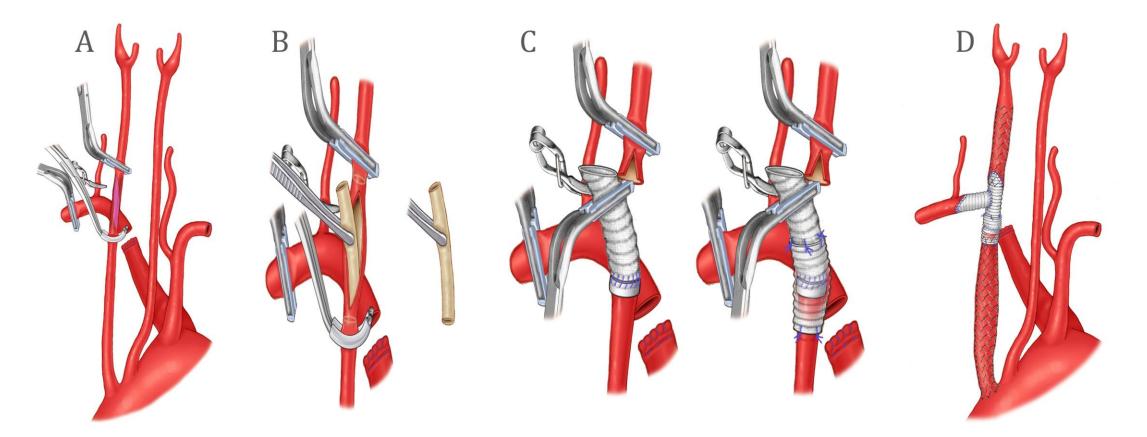


Figure 3. Right carotid dissection. Interposition grafts. Right carotid- subclavian bypass. Carotid stents.



- The 2nd stage was performed with left thoracotomy (muscle-sparing).
- The proximal Kommerell diverticulum was clamped and controlled. Using multiple 3-0 Prolene pledget sutures, the diverticulum was ligated and resected.
- Patient was discharged home stable on postoperative day 7.







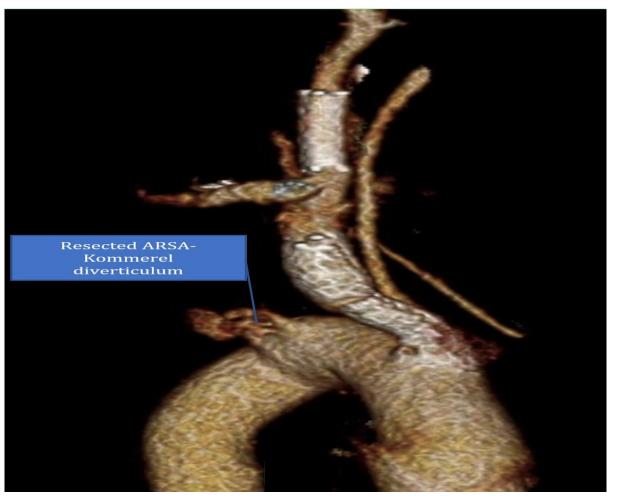
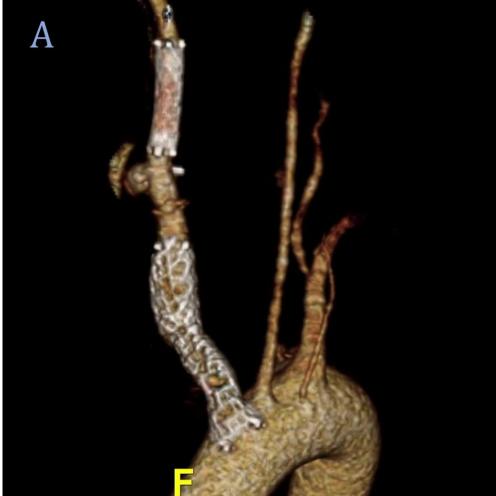


Figure 4. Patent R carotid-subclavian bypass, RCCA stent, and ligation - resection of the origin of ARSA and Kommerell diverticulum.





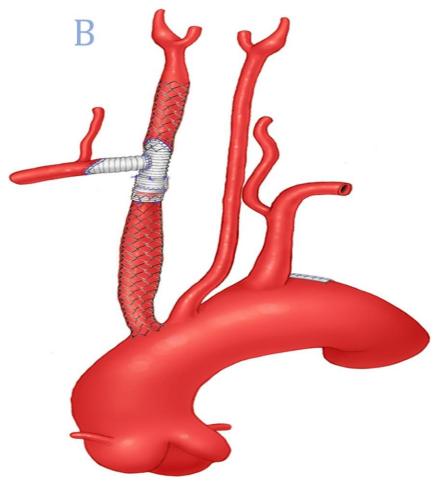


Figure 5. Postoperative 3D reconstruction revealing patency of the innominate and RCCA stents (Figure 5A). Final outcome reconstruction (Figure 5B).



DISCUSSION

- Up to 60% of patients with aberrant right subclavian artery (ARSA) also exhibit a wider origin of the vessel, referred to as Kommerell's Diverticulum (KD).
- Our case illustrates the **potential complications** that may arise when addressing supra-aortic or subclavian arteries in patients with Loeys-Dietz syndrome (LDS), particularly in the complex region of the proximal right subclavian artery and innominate artery.

DISCUSSION

- In patients with LDS, characterized by **fragile vascular integrity**, it is essential to ensure appropriate management of arterial complications during surgery. This involves specific techniques
- Management of arterial complications:
 - Interrupted sutures.
 - Preventing potential tears or dissections.
 - Protecting the clamps (Plasma tubing or Fogarty hydro-grip clamps).



Giuliani L. J Am Coll Cardiol. 2023;81(10):979-991.



CONCLUSIONS

The presented case highlights the importance of a multidisciplinary approach and **meticulous surgical planning** when treating dysphagia lusoria in patients with underlying connective tissue disorders such as Loeys-Dietz syndrome.



