



## Mediastinoscopy-assisted Treatment of an Aberrant Right Subclavian Artery

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Aberrant right subclavian artery (ARSA) is a rare congenital anomaly characterized by the origin of the right subclavian artery from the aortic arch distally to the left subclavian artery. We describe the case of a young patient with symptomatic ARSA treated by mediastinoscopy-assisted ligation at its origin and subclavian—carotid transposition.

The aberrant right subclavian artery (ARSA) is a rare congenital anomaly characterized by a retroesophageal course of the right subclavian artery, whose origin from the aortic arch is distal to the left subclavian artery. The majority of patients with ARSA are asymptomatic; however, this anomaly may cause a condition named *dysphagia lusoria*.

## **CASE REPORT**

A 36-year-old woman was referred to our center for a history of progressively worsening dysphagia and severe loss of weight. The patient reported an initial dysphagia for solids which had started 2 months prior. This has recently progressed to a dysphagia for liquids as well. Both neurological and otolaryngology evaluations were negative for organic or functional swallowing impairments. Due to her malnourished conditions, parental nutrition was administered. Videofluoroscopy was then performed and demonstrated an external compression during bolus progression in the proximal zone of esophagus on the

anterolateral left side of the esophagus. This finding was considered highly suggestive for an ARSA. A computed tomography angiography (CTA) confirmed the diagnosis of an 8-mm ARSA and also revealed the associated anomalous origin of the left common carotid artery from the innominate trunk (Fig. 1). Due to the significant symptoms, surgical intervention was then planned. Under general anesthesia, the patient was placed in the supine position. A sandbag was positioned in the midline behind her shoulders to achieve a better extension of the neck. Through a 6 cm right supraclavicular incision, we sectioned the clavicular head of the sternocleidomastoid muscle and the anterior scalene muscle, after identification of the phrenic nerve. This helped to expose the ARSA, the mammary artery, the thyreocervical trunk, and the right vertebral artery. Then, with the use of a mediastinoscope, we followed the proximal aberrant subclavian artery in the retroesophageal space until 1 cm from its origin off the aortic arch. After systemic heparinization, a curved Satinsky vascular clamp was placed at the origin of ARSA (Fig. 2A); then, we oversewed the proximal portion of this artery as close to its origin as possible with a 2 Mersilene suture and transected it. The distal portion of the right subclavian artery was trimmed and with a careful preservation of the right vertebral artery, an end-to-side anastomosis was made with the right common carotid artery with a polypropylene 6-0 (Fig. 2B). No intraoperative complications occurred. Her postoperative course was uneventful and she tolerated regular diet with immediate regression of her dysphagia. On second postoperative day, a Gastrografin transit confirmed the absence of any obstruction in the esophageal progression. On the fourth postoperative day, a CTA was done to confirm the good result of the repair. She was discharged 7 days after surgery in good clinical condition, with palpable radial pulses and no neurological signs. Currently, after a follow-up of 11

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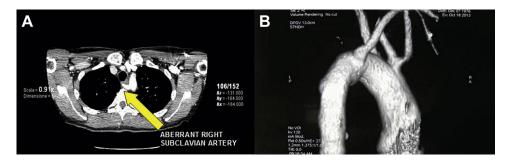
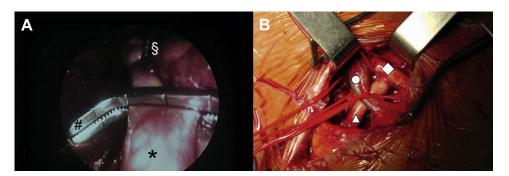


Fig. 1. (A) Preoperative CTA axial scan. (B) Posterior three-dimensional view showing a truncus bicaroticus and the ARSA.



**Fig. 2. (A)** Mediastinoscopy-assisted clamping at the origin of the vessel from the aortic arch. **(B)** Subclavian—carotid transposition. #: curved Satinsky clamp; \*:

distal ARSA;  $\S$ : Mersilene ligation of the proximal ARSA;  $\bigcirc$ : right internal jugular vein;  $\diamondsuit$ : right common carotid artery;  $\Delta$ : aberrant right subclavian artery.

months, the patient is still asymptomatic and the success of the procedure has been confirmed by a CTA at 1 and 6 months (Fig. 3).

## **DISCUSSION**

The ARSA is the most common embryological abnormality of the aortic arch and it occurs in 0.5—1.8% of the population. In 1794, Bayford described the association between the postmortem diagnosis of an ARSA with patients' symptoms and named this condition "dysphagia lusoria" from *lusus naturae*, like a freak of nature. <sup>2</sup>

Embryologically, the proximal part of the right subclavian artery normally originates from the fourth right aortic arch. When this arch does not evolve in the correct way during embryological development, the persistence of the distal right dorsal aorta leads to the formation of an aberrant origin of the right subclavian artery distal to the left subclavian artery. Commonly, this vessel passes behind the esophagus and rarely between the esophagus and the trachea or anterior to both of the

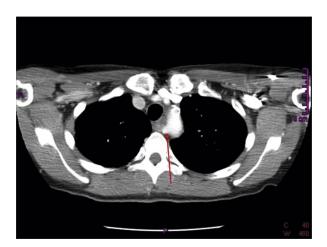


Fig. 3. Postoperative CTA (red arrow: ARSA stump).

structures.<sup>3</sup> When the origin of the vessel has sufficient dorsal aortic remnants, there is a condition called Kommerell diverticulum (KOD), an anomaly described for the first time in 1936 by Burckhard Friedrich Kommerell.<sup>4</sup> Although the majority of patients affected by ARSA with or without KOD are

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asymptomatic, esophageal compression can cause difficulty in swallowing. Other symptoms described are interscapular pain, cough, hematemesis, and dyspnea.<sup>1</sup>

Barium contrast examination of the esophagus is a good diagnostic tool, demonstrating a characteristic impression defect at the level of the third and fourth vertebra<sup>5</sup>; CTA and/or angiography usually are the best examinations to confirm the diagnosis.<sup>5</sup>

Due to the rarity of this condition, indications for surgical intervention have been described, but not fully agreed upon. These include symptomatic patients with an ARSA with or without KOD, or patients in whom the KOD aneurysmal degeneration is >3 cm.<sup>3</sup>

The first surgical approach was performed by Gross in 1946: through a left thoracotomy he simply ligated and divided the aberrant artery. Since then, several other approaches have been described, such as right thoracotomy, sternotomy, cervical and supraclavicular access.

During recent years, different endovascular and hybrid techniques have been proposed: distal ligation of the ARSA and its endovascular occlusion, associated with or without right carotid—subclavian bypass or right subclavian—carotid transposition, exclusion with endoprosthesis alone, or insertion of a covered stent into ARSA.

Unfortunately, long-term outcome data are not available and these techniques have a reported post-operative complication rate of 22% and endoleak rate of 13%.<sup>3</sup> This was the reason why an endovascular treatment was not considered for our patient (36 years old).

As far as we know open ligation and right carotid artery revascularization through a supraclavicular approach is the treatment with the lowest associated morbidity. The most challenging part of this approach is the exposure of the proximal portion of the ARSA as close to its origin as possible, to avoid any possible aneurysmal degeneration of a residual

stump. The use of mediastinoscope turned out to be helpful for this purpose, because we could easily identify the origin of the aberrant vessel. This is a delicate maneuver and has to be performed by a skilled and experienced thoracic surgeon, as we did; in fact, a wrong use of this instrument may cause iatrogenic esophageal, tracheal, or vascular lesions. In 2009, Moorjani<sup>7</sup> described this technique for the first time and in 2012 Fukuhara<sup>8</sup> confirmed its effectiveness.

To our knowledge, this is the third case of a cervical approach to the ARSA assisted by mediastinoscopy. We believe that endovascular/hybrid options could be a reasonable alternative in patients affected by an ARSA with KOD or aneurysmal degeneration, whereas isolated cases of ARSA could be easily treated through mediastinoscopy-assisted cervical approach.

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