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## Diverticulum of Kommerell

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### Summary

A case of an 11-months-old girl with vomiting and laryngeal stridor is presented, and in whom a Kommerell diverticulum was demonstrated, which is a rare variant of the incomplete vascular ring. It is well known that the magnetic resonance is the best study to define this malformation. In this patient, it was the cardiac catheterization which allowed to define the structures that constituted the vascular ring. The patient was surgically treated in a successful way. The embryological knowledge of the transformations of the aortic arches offers great assistance in the understanding of all the types of vascular rings.

**Key words:** Kommerell's diverticulum. Vascular rings. Dysphagia.

**Palabras clave:** Divertículo de Kommerell. Anillo vascular. Disfagia.

### Resumen

#### DIVERTÍCULO DE KOMMERELL

Se presenta el caso de lactante de 11 meses con historia de vómitos y estridor laríngeo secundaria a una rara variante de anillo vascular, divertículo de Kommerell. La resonancia magnética es considerada como el estudio ideal para definir la malformación, sin embargo, en este paciente, fue la angiografía la que permitió definir las estructuras del anillo vascular. El paciente fue tratado quirúrgicamente de manera exitosa. El conocimiento embriológico permite un adecuado entendimiento de los diferentes tipos de anillo vascular.

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### Introduction

**T**he term vascular ring is referred to the alterations of the aortic arches in which the trachea and the esophagus are surrounded by these structures. They are divided in complete and partial; the former are the double aortic arch and the latter come from the aberrant origin of a subclavian artery, or arterial ligament or duct, counterside to the aortic arch. The symptoms and more characteristic signs are: persistent stridor and difficult feedings. The moment of the presentation is varied, but it normally occurs in the neonatal period.<sup>1,2</sup> The purpose of this article is to present the case of a rare variant of the vascular ring with right aortic arch.

### Presentation of the case

Female patient, 11 months old, referred to our Institute for vomits and laryngeal stridor occurring since the neonatal age. A barium esophagogram was carried out which showed right-sided indentation of the esophagus (*Fig. 1*). The echocardiogram did not find any intracardiac defect. The magnetic resonance did not defined accurately the structures which formed the vascular ring. The angiography showed right aortic arch from which the arteries emerged in this order: left brachycephalic arterial trunk situated on the left, right carotid artery and right subclavian artery. Left subclavian artery was pull by a ligament which began from a retroesophagic diverticulum from the descending aorta (*Figs. 2 - 3*).

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**Fig. 1.** Barium esophagogram showing indentation of esophagus by aortic arch anomaly.

Surgical resection of the diverticulum, and section of the ligament was done so disappearing the digestive, and respiratory symptoms.

### Discussion

The vascular rings represent less than 1% of all the cardiovascular malformations.<sup>1,2</sup> This malformation is established for the persistence or the loss of specific segments from the rudimen-

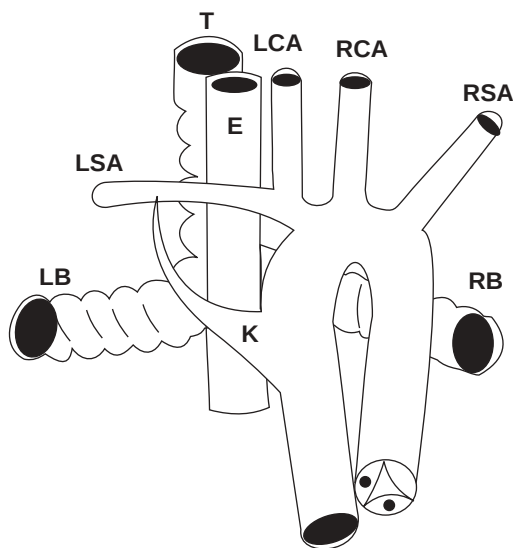


**Fig. 2.** Aortography demonstrating diverticulum of Kommerell. **K:** Kommerell's diverticulum; **LSA:** left subclavian artery; **LA:** ligamentum arteriosum; **Ao:** Right-sided aortic arch.

tary aortic arch. In the embryology of the aortic arch system, around the 36 to 38 days, six pairs of branchial arches originate six aortic arches with left-right symmetry, so constitute the primitive vascular back up of the brachycephalic structures.<sup>3,4</sup>

The embryonic aortic arches system are bilateral vessels which connect the ventral and dorsal aortas, this produces a bilateral vascular system that surrounds the fore gut intestine from which the trachea and the esophagus come from.

The distal parts of the first and second arches disappear close to the dorsal aorta, and the proximal areas of them close to the ventral aorta keep on as hyoid and maxillary arteries respectively, which are branches of the external carotid artery. The third aortic arches persist and form the proxi-



**Fig. 3.** Diagram showing vascular ring in posterior view. **K:** Kommerell's diverticulum; **E:** Esophagus; **T:** Trachea. **LB:** Left bronchus; **RB:** Right bronchus; **LCA:** Left carotid artery; **RCA:** Right carotid artery; **LFA:** Left subclavian artery; **RSA:** Right subclavian artery.

mal parts of the internal carotid arteries. On the other side the dorsal aorta between the third and fourth aortic arches known as carotid ducts, involute too. The fourth aortic arches persist encircling the anterior gut, this symmetry gives up an asymmetric pattern when one segment of the right dorsal aorta normally involutes between the seventh segmental artery and the place of union of both dorsal aortas which determines the formation of the aortic arch to the left. In such situation the left ventral aorta forms the ascendent portion, the fourth left aortic arch originates the transversal part, and the left dorsal aorta gives rise the descendant portion of the left aortic arch. The right ventral aorta give origin to the brachycephalic arterial trunk, from which the right common carotid artery and the

right subclavian artery emerge, this last one has a tripartite origin, the proximal part derives from the right fourth aortic arch, the intermediate segment from the right dorsal aorta between this arch and the origin of the seventh segmentary artery, this last one constitutes the distal part.

The sixth aortic arches originate the proximal portions of the branches of the pulmonary artery and the right and left arterial ducts, the right one usually disappears and the left one keeps or remain patent during fetal life.

The Kommerell's diverticulum represents the persistency of the distal segment of double aortic arch, generally the left one which the proximal segment is atretic or disappears. Usually it is not associated with other congenital heart disease, some authors have reported the ventricular septal defect as the most frequent malformation, other series mention aortic coartation, tetralogy of Fallot and transposition of the great arteries.<sup>1-5,7</sup>

The clinical manifestations of the vascular ring and the seriousness will depend directly on the compression degree, in most of the patients the symptoms will appear before one month old, among the most known are the persistent stridor and feeding difficulty.<sup>2,6,7</sup>

The magnetic resonance is the best noninvasive techniques to demonstrate the anatomic features of vascular ring and thus to determine the optimal surgical approach. The cardiac catheterization is indicated when there is a confusion over the structures that form the vascular ring as it was in the case herewith.

The surgical treatment is indicated in patients with symptoms of airway or esophageal compression. Ciná reported a surgical mortality of 8.3% for elective treatment of Kommerell's aneurysm and Austin reported that 19% of affected patients presented with rupture, and all of them died.<sup>9-13</sup>

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