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# Kommerell Diverticulum: Right Aortic Arch with Anomalous Origin of Left Subclavian Artery and Duplicity of Right Vertebral Artery in a 16-Year-Old Girl

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Data Collection B  
Statistical Analysis C  
Data Interpretation D  
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**Conflict of interest:** None declared

**Patient:** Female, 16  
**Final Diagnosis:** Kommerell diverticulum  
**Symptoms:** Tachycardia  
**Medication:** —  
**Clinical Procedure:** Clinical observation only  
**Specialty:** Cardiology





**Objective:** Rare disease  
**Background:** Kommerell diverticulum with a right-sided aortic arch is a rare congenital anatomical condition most often observed in adults. A possible etiology of the subclavian artery's anomalous origin would be an abnormality in regression of the fourth primitive aortic arch during embryonic development.

**Case Report:** We report on the case of a 16-year-old female patient presenting with complaints of occasional tachycardia and mild non-specific dyspnea after anxiety crises. Physical examination revealed lower amplitude of the pulses in the left upper limb compared to the right upper limb, and difference in blood pressure (BP) values of 80×60 mmHg, and 100×60 mmHg, respectively. Different radiological imaging modalities were performed to elucidate a possible vascular abnormality. Multislice detector computed tomography angiography of the thoracic aorta and supra-aortic trunks showed a right-sided aortic arch and an aberrant origin of the left subclavian artery with a retroesophageal course and dilation of its emergence (Kommerell diverticulum), as well as duplicity of the right vertebral artery (RVA). Considering the actual small diameter of the diverticulum and the absence of dysphagia or severe external esophageal compression analyzed by the esophagogram, vascular surgery was not indicated. Since complications have been described in the literature, the patient must be kept under observation in the future.

**Conclusions:** Congenital vascular alterations, including Kommerell diverticulum with right-sided aortic arch and the aberrant origin of the left subclavian artery, should be suspected in otherwise asymptomatic young patients with few clinical manifestations. Investigation with different imaging methods helps to clarify the vascular abnormalities, to support a possible surgical procedure indication, and to monitor the patients in follow-up.

**MeSH Keywords:** Angiography • Aorta, Thoracic • Deglutition Disorders • Diverticulum • Subclavian Artery

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## Background

Kommerell diverticulum with a right-sided aortic arch is a rare congenital anatomical condition most often observed in adults. It can result from dilation of the anomalous origin of the subclavian artery in the following situations: left aortic arch (LAA) with anomalous origin of right subclavian artery (RSCA) (0.5–2.0%), or right aortic arch (RAA) with anomalous origin of the left subclavian artery (LSCA) (0.05–0.1%) [1,2].

A possible etiology of the subclavian artery's anomalous origin is an abnormality in regression of the fourth primitive aortic arch during embryonic development [3]. This condition was named Kommerell diverticulum in honor of Dr. Kommerell, the radiologist who first made the diagnosis [4]. Anatomical changes are usually not associated with clinical symptoms, and the diagnosis of Kommerell diverticulum typically occurs incidentally during adulthood [3]. In some situations, especially when there is a right aortic arch and left subclavian artery, the diverticulum may be associated with dysphagia and dyspnea due to tracheal and/or esophageal compression, which directly influence the clinical and surgical treatment of this pathology. This case report demonstrates a variant of Kommerell diverticulum with a right aortic arch and aberrant left subclavian artery, associated with another vascular congenital anatomic variation – the duplicity of the right vertebral artery (RVA).

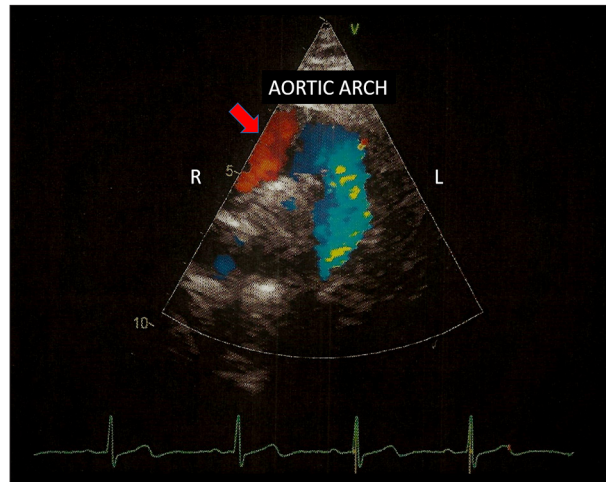
## Case Report

We report the case of a 16-year-old female patient presenting with complaints of occasional tachycardia and mild non-specific dyspnea after anxiety crises. Physical examination revealed lower amplitude of the pulses in the left upper limb (LUL) compared to the right upper limb (RUL), and a difference in blood pressure (BP) values: BP RUL 100×60 mmHg and BP LUL 80×60 mmHg.

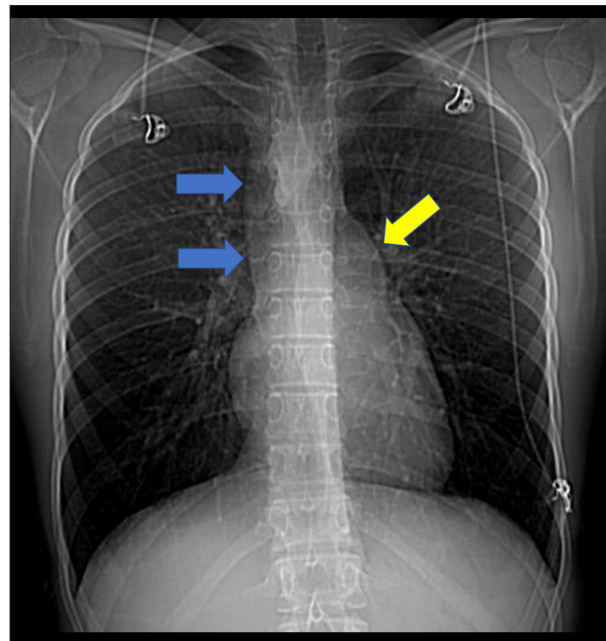
The electrocardiogram was within normal range, and 24-h Holter monitoring showed no arrhythmia.

The transthoracic color Doppler echocardiography (Echo) revealed a right aortic arch (Figure 1) with emergence of a vascular structure, and a three-phase pulsed pattern flow in the infra-aortic region on the right, suggestive of an aberrant subclavian artery.

A 16-multislice detector computed tomography angiography (MDCTA) of the thoracic aorta and supra-aortic trunks was obtained using standard protocols with 1-mm collimation during the venous administration of iodinated nonionic contrast. This was followed by multiplanar and three-dimensional reconstructions using multiplanar reconstruction (MPR) and



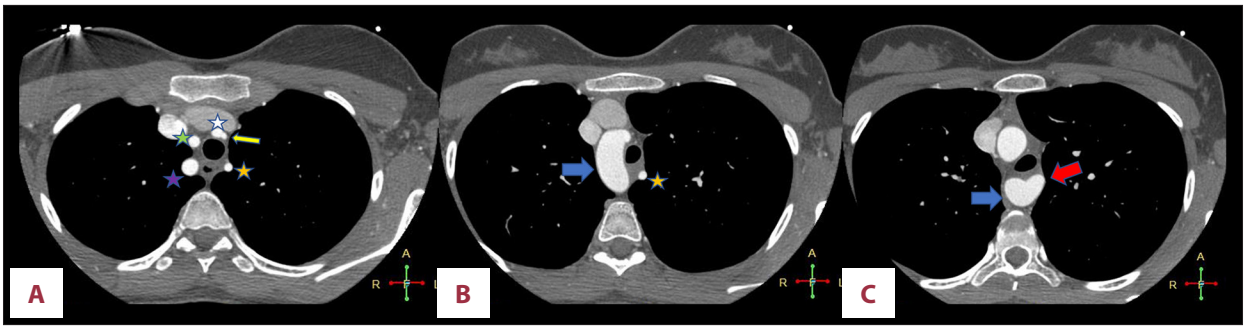
**Figure 1.** Color Doppler echocardiography showing right aortic arch.



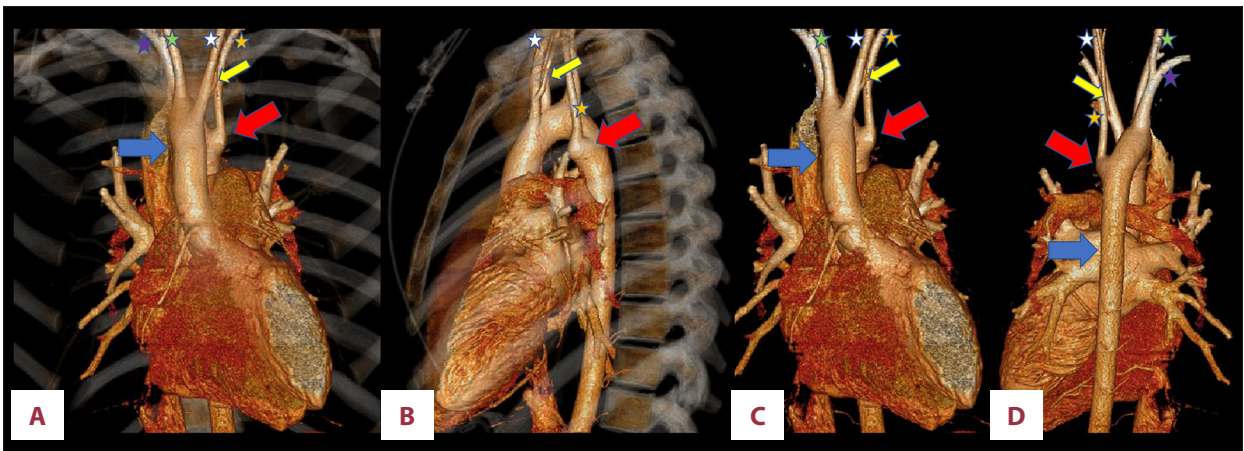
**Figure 2.** The frontal digital topogram of the MDCT shows the normal position of the pulmonary artery trunk (yellow arrow) and enlargement of the right upper mediastinum, caused by the sum of the RAA and superior vena cava (blue arrows).

volume-rendering techniques (VRT) (Figures 2–4), which demonstrated the presence of RAA, causing a mild impression on the right lateral border and a mild leftward displacement of the trachea. MDCTA also showed the aberrant origin of the LSCA, with a retroesophageal course and dilation of its emergence – Kommerell diverticulum – as well as duplicity of RVA.

The patient underwent upper digestive endoscopy, which showed a slight extrinsic compression in the middle third of



**Figure 3.** Axial MDCT images of the upper mediastinum show in **A**, the origin of the LVA (open red arrow) is observed in the ipsilateral common carotid (open black arrow), the position of the right common carotid artery (open blue arrow), the RSCA (open green arrow), and the LSCA (open yellow arrow). In **B**, images show the RAA (solid blue arrows), and in **C**, Kommerell diverticulum (solid red arrow) at the level of the origin of the LSCA.



**Figure 4.** (A–D) MDCT images with three-dimensional reconstruction of the thoracic aorta using VRT (Volume Rendering Technique) – **A** and **C** coronal views, **B** left sagittal view and **D** posterior view show the heart, thoracic aorta and the origin of the supra-aortic trunks. **A** and **B** also show the thoracic bones. The images show RAA (blue solid arrows in **A**, **C** and **D**), anatomical condition involving the origin of the supra-aortic trunks (white open arrow – left common carotid artery in **A–D**, blue open arrow – right common carotid artery in **A**, **C**, **D**, red open arrow – RSCA and yellow open arrow – LSCA in **A–D**) and Kommerell's diverticulum (red solid arrow) at the level of the origin of the LSCA in **A–D**. Origin of the LVA (yellow solid arrow) in the ipsilateral common carotid artery is shown in **A–D**.

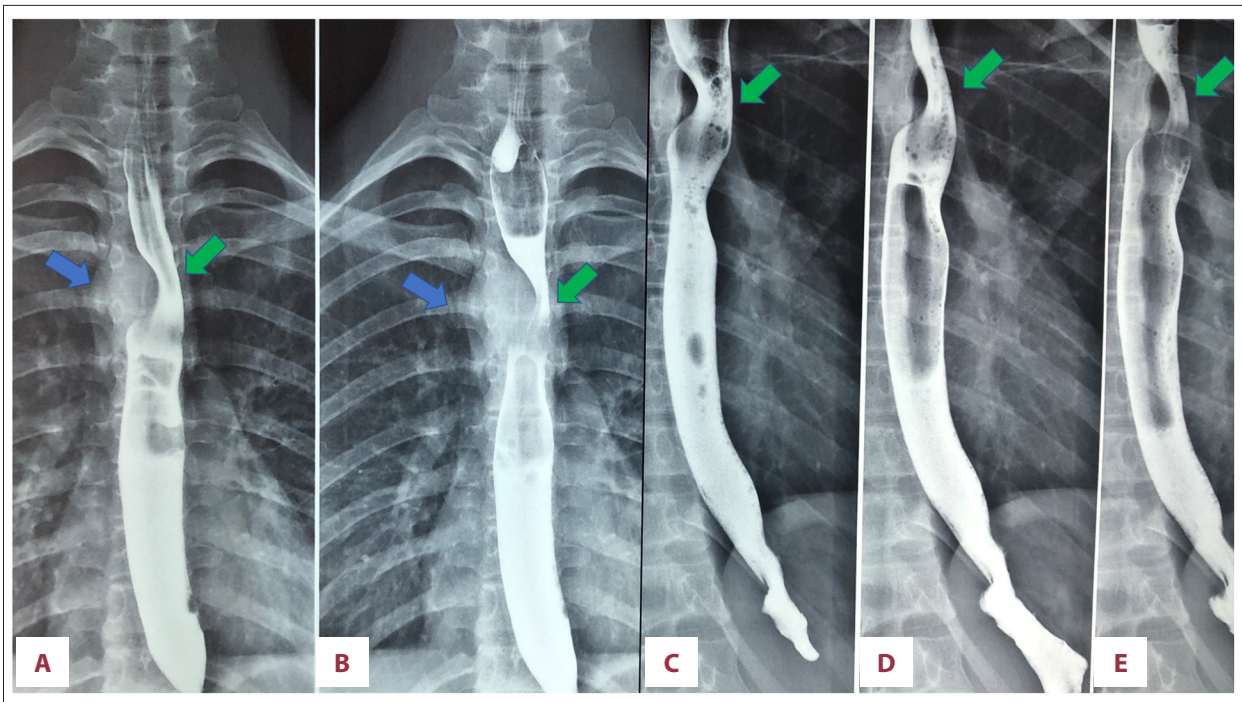
the esophagus, and a contrast radiographic study of the esophagus (barium esophagram) that showed a discrete esophageal impression by the RAA (Figure 5).

Vascular surgery was not indicated considering the actual diameter of the diverticulum and the absence of dysphagia or severe external esophageal compression shown by the esophagogram. Since complications have been described in the literature, the patient was kept under observation. There was spontaneous disappearance of dyspnea and tachycardia, and no new symptoms appeared. The patient remains without medication and is in clinical follow-up.

## Discussion

Kommerell diverticulum can be defined as a bulb-like swelling of the proximal portion of an aberrant left subclavian artery, adjacent to its aortic origin. It is a rare condition, which can occur with either the LAA and RSCA aberrant origin (0.5–2% of the population) or with the RAA and LSCA aberrant origin (0.05–0.1% of the population) [1].

One of the classifications described in the literature reports that the aortic diverticulum is located at the aorto-ductal junction, perceived as a bulge along the internal portion of the aortic isthmus distal to the LSCA [3]. Another classification describes the persistent proximal portion of the dorsal aorta as the result of the formation of a Kommerell diverticulum [5].



**Figure 5.** Barium esophagram. The images show RAA (blue arrows) and the impression caused in the esophagus (green arrows) during swallowing of the barium contrast at different moments. **A** and **B** frontal views, and **C-E** right lateral views.

Literature reviews have found Kommerell diverticulum to be present in up to 100% of patients with a RAA and an aberrant LSCA diagnosed by routine CT examination, and about 60% of patients with an aberrant subclavian artery, the most common aberrant artery on the left side, have a Kommerell diverticulum [6]. The location of the diverticulum is posterior to the esophagus in 80% of cases, between the trachea and the esophagus in 15% of cases, and anterior to the trachea in 5% of cases [7]. One explanation for the anomalous origin of the RAA is given by the diagram of Rathke's pouch, which consists of an embryological malformation with remodeling of the 6<sup>th</sup> ventral and dorsal arches during the 4<sup>th</sup> and 5<sup>th</sup> weeks of embryonic life [8].

The patient in this report presented with an RAA from an aberrant LSCA origin. These genetic abnormalities are usually discovered incidentally during routine imaging tests, but it is very important to diagnose the symptomatic forms early. RAA is diagnosed in about 0.1% of the general population, and about 50% of these individuals have an aberrant LSCA origin [9]. An abnormal LSCA origin can occur without any concurrent anomaly; however, it is more commonly associated with RAA [10].

Some authors report that an RAA without a simultaneous congenital heart defect is extremely rare [11]. In 5–10% of cases, it is related to congenital heart diseases, including tetralogy of Fallot, pulmonary stenosis with ventricular septal defect, tricuspid atresia, and patent ductus arteriosus [11]. In patients

with these anomalies, the aberrant subclavian arteries usually have a course posterior to the esophagus towards the contralateral upper limb, but our patient did not have cardiac congenital abnormalities associated with RAA and aberrant LSCA. Other authors have reported that RAA with an aberrant LSCA should always have a left ductus arteriosus that can form a vascular ring connecting the left pulmonary artery to the aberrant LSCA origin [1]. This vascular ring causes tracheal compression, and when there is no compression or tracheal deformity, the ring may not be present; if it is, it does not need to be corrected [1,5].

Most patients do not show any symptoms, and the diagnosis is made incidentally in imaging studies obtained for other non-related reasons [5]. Progressive symptoms related to compression of the esophagus (dysphagia) or trachea (stridor) may be observed in cases with large dilatations or with localized diverticula at specific sites [6]. Cardiovascular symptoms can also occur as a result of associated structural cardiac malformations, early atherosclerotic lesions, or abnormal artery dissection [5]. Our patient's complaints were mostly associated with anxiety episodes, and they disappeared spontaneously during the initial follow-up. Thus, after evaluation with specific tests, they were considered unrelated to structural vascular disease.

The only physical sign found in this patient was a discreet difference in pulse and systolic BP between the RUL and LUL, which may be related to the aberrant origin of the LSCA. With

the supine position of the LUL, the absence of arterial pulse has been described previously [3]. Although our patient presented with RAA, causing a mild impression on the trachea and an aberrant LSCA with a retroesophageal course, she had no symptoms such as dysphagia, chest pain, or stridor.

The key surgical indication occurs when the Kommerell diverticulum presents with a diameter greater than 5 cm in symptomatic patients; in such a situation, complications such as tracheomalacia, esophageal dilation, or rupture of the aneurysm may occur [12]. As the diverticulum of our patient was 15×13 mm in diameter, it was considered small, and the medical decision was to maintain periodic clinical follow-up.

Abnormal findings were observed by transthoracic Echo, MDCT of the thoracic aorta, and contrasted radiographic esophageal study of the patient. These tests were important for a comprehensive understanding of the vascular anatomy and its relationships with adjacent structures, as well as to verify concomitant cardiac structural alterations.

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## Conclusions

Congenital vascular alterations, including Kommerell diverticulum with right-sided aortic arch and the aberrant origin of the left subclavian artery, may be suspected in otherwise asymptomatic young patients with few clinical manifestations. Investigation with different imaging methods helps to clarify the vascular abnormalities, to support a possible surgical procedure indication, and to monitor the follow-up of the patients.