# Dysphagia lusoria caused by a right-sided aorta

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## Abstract

**We report a case of an 80-year-old female with dysphagia lusoria caused by oesophageal compression by a right-sided atheromatous aorta. The relationship between aortic root vascular anomalies and dysphagia has been clearly established in literature and can be diagnosed by a barium swallowing study, followed by CT or MRI. Aortic anomalies and variations in aortic branches are caused by embryonic malformations and are mostly described in association with congenital heart lesions. In this pauci-symptomatic patient, the preferred treatment is a conservative management.** (Acta gastroenterol. belg., **2012**, 75, **266-269**).

**Key words** : dysphagia lusoria, vascular anomalies, right-sided aorta.

## Introduction

A right-sided aorta is a congenital anomaly that is very rare and often asymptomatic. Symptoms occur when the aorta-or an aortic branch-causes compression on other mediastinal structures, like the oesophagus or the trachea. Our patient presented only at the age of 80. The reason for this late presentation is the age-related atheromatous transformation of the aorta, causing compression on the oesophagus.

## Case report

An 80-year-old female presents to our department of gastroenterology because of episodes of choking with persistent cough. Her medical history includes diabetes mellitus (treated by oral antidiabetics), hypercholesterolemia (treated by a statin) and, since childhood, a central paresis of the facial nerve of unknown cause. A few months ago she underwent an echocardiography showing basal septal hypertrophy with systolic anterior movement of the mitral valve with moderate insufficiency of the mitral valve. Her cardiologist prescribed her a beta blocker.

History reveals prominent dysphagia for solids and fluids, with choking for two months. She has no complaints of vomiting and there is slight nausea. She mentions anorexia for the last year, but is not aware of the presence of any weight loss. Systemic anamnesis reveals no other remarkable abnormalities.

Physical examination reveals a lean patient with normal cardiopulmonary and abdominal examination. There are no palpable adenopathies.

Laboratory results show a normochromic normocytary anemia with a hemoglobin level of 11,6 g/dL (12,0-16,0) with a normal iron an vitamin status. Other results are a slight trombopenia of 123 × 10^9/L (145-367), discrete elevation of gamma GT (gamma glutamyl transpeptidase) to 77 U/L (12-43) and GOT (glutamate oxaloacetate transaminase) to 70 U/L (0-59). Inflammatory parameters are within normal limits.

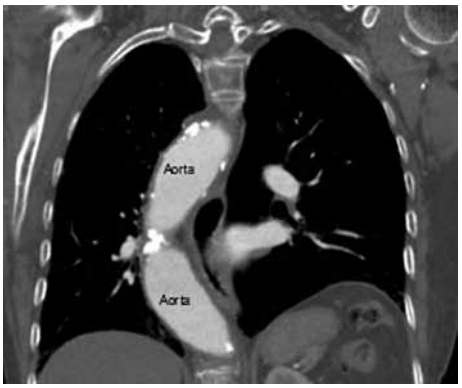


Fig. 1. — Coronal CT-images showing the right-sided atheromatous descending aorta (aorta).

A gastroscopy is performed and appears to be normal. On chest radiograph an aneurysmal dilation of the aortic arch is suspected with an estimated diameter of 53 millimeters. Because of this finding, computed tomography of the chest is performed, which shows a right-sided aortic arch with a descending aorta on the right side of the spinal column (Fig. 1). In addition there is an atheromatous dilation of the entire descending aorta, without an aneurysmal dilation. A surprising finding is the absence of a brachiocephalic trunk on the right side, where the subclavian artery and carotid artery have a separate origin. Moreover a diverticulum at the origin of the left subclavian artery is noticed, which is known as a Kommerell's diverticulum.

An otopharyngolaryngeal consultation shows a normal mouth and throat with normal function of the tongue. Laryngotracheoscopy performed by the otopharyngolaryngist confirms that the patient easily chokes when

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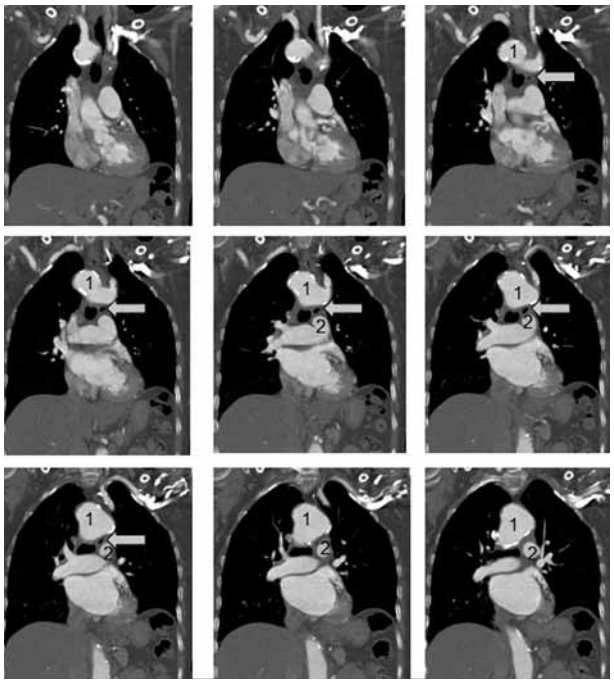


Fig. 2. — Coronal CT-images, that show progressive narrowing to compression of the oesophagus (bold arrow) between the atheromatous aorta (number 1) and the pulmonary artery (number 2).

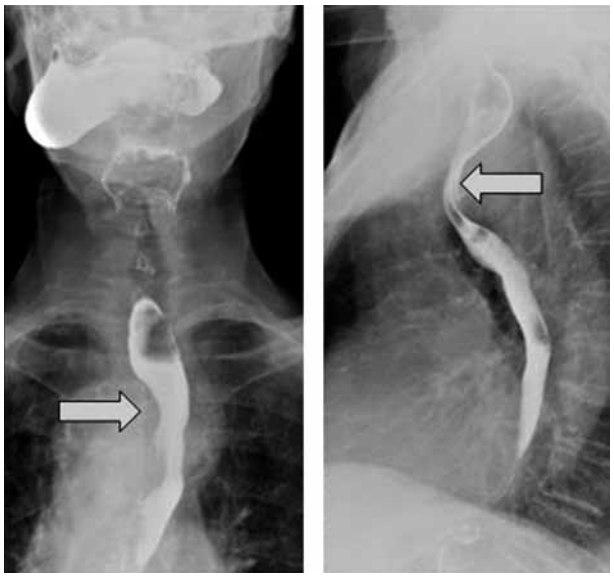

Fig. 3. — Images of the videofluoroscopic swallowing study show an obvious extrinsic compression of the oesophagus by the aortic arch, with deviation to the left and anterior side (bold arrows).

drinking water, but they see no stasis of fluids and a good coughing reflex.

A videofluoroscopic swallowing study shows an important extrinsic imprinting on the high mediastinal oesophagus, caused by the above mentioned right-sided aortic arch, that compresses the oesophagus between the pulmonal artery and the aortic arch (Fig. 2, Fig. 3). This causes a deviation of the oesophagus to the left and anterior side with imprinting on the right posterior lining of the oesophagus.

We concluded that the dysphagia in this patient was caused by the oesophageal compression by the atheromatous right-sided aortic arch. We reassured our patient and decided not to perform any surgical interventions because of the benign character of our findings with only mild symptoms and we proposed slight dietary modifications.

An oesophageal manometry was not performed, but we have no arguments to assume a functional origin in this patient with a history compatible with mechanical dysphagia.

## Discussion

*Relationship between dysphagia and vascular anomalies*

The syndrome of oesophageal compression by an abnormal vascular structure was first described by Bayford in 1794, following a case of dysphagia caused by compression of the oesophagus by an anomalous right subclavian artery, that was diagnosed on autopsy (1).

External compression of the oesophagus by a vascular structure is an uncommon cause of dysphagia. Case reports of almost every major vasculature structure within the chest cavity causing some degree of oesophageal compression and symptomatic dysphagia have been described in literature (2). It requires a high index of suspicion to make the diagnosis of dysphagia lusoria, which is nowadays commonly described in literature as 'difficulty in swallowing caused by an aortic root vascular anomaly'.

*Vascular anomalies of the aortic arch*

Developmental anomalies of the aortic arch and its major branches are relatively common, being found in 3% of people in autopsy studies. The most common congenital aortic anomaly is an aberrant right subclavian artery (2). A right-sided aorta was first described in 1763 by Fioratti and Aglietti (3). This is a very rare condition that only occurs in 0,05% to 0,1% of radiology series and in 0,04% to 0,1% of autopsy series (4). A right-sided aorta is mostly associated with congenital heart lesions like Tetralogy of Fallot, ventricular septal defects, pulmonary stenosis, truncus arteriosus or transposition of the great vessels (5) and is rare in the presence of a structural normal heart (4). In our case report we have no arguments for congenital cardiac lesions, although we have to mention that a recent echocardiography a few months before admission showed a basal hypertrophy of the septum with a systolic anterior movement of the mitral valve. Beta blockade was started and a new echocardiography a few months afterwards no longer showed a systolic anterior movement of the mitral valve.

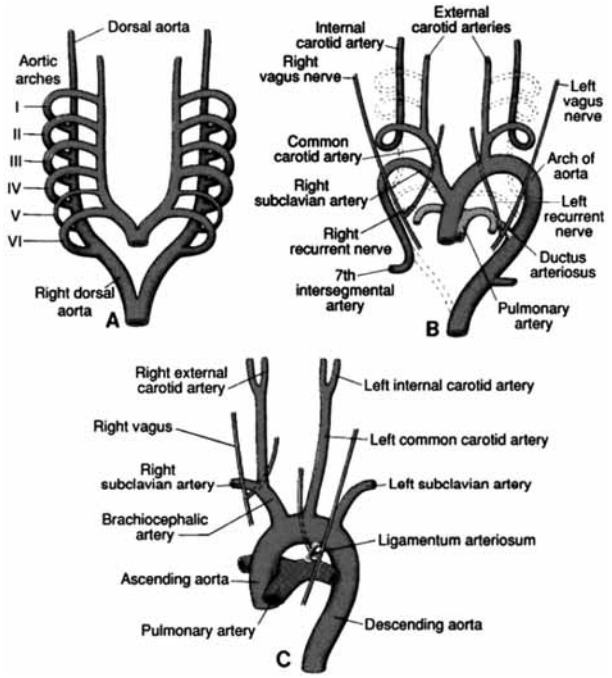


Fig. 4. — Normal embryonal development of the great vessels. (A) In early in-utero development, the aortic arches are a bilateral duplicate system. (B) As the great vessels develop, the right aortic system undergoes atresia. (C) Fully developed anatomy of the great vessels at birth (2).

A right-sided aortic arch is a congenital anatomical anomaly that has its origin in an aberrant organogenesis. In the embryo, six pairs of aortic arches develop at different stages of the organogenesis. The fourth primitive left aortic arch will form the adult aortic arch. The right fourth generally disappears, producing the normal course of the aorta arching to the left and descending to the left of the spine (Fig. 4). If both arches persist, they form a double arch or a vascular ring encircling the trachea and oesophagus. If the left aortic arch disappears and the right one persists, a right-sided aortic arch is formed (4,6). The last anomaly is mostly associated with an aberrant origin or an abnormal course of the aortic branches (5). The etiology of this embryonic abnormity is not known.

Fifty percent of right-sided aortas are associated with a Kommerell's diverticulum or aneurysm. This anomaly was indeed present in the case presented above. This entity is present when the subclavian artery has its origin in a diverticulum of the aortic arch and was first described by Kommerell in 1936 (4,5,7).

*Symptoms of vascular anomalies*

Vascular anomalies of the aortic root are in most cases completely asymptomatic.

Infants mostly present with respiratory symptoms, like dyspnea, wheezing or recurrent pneumonia (2,4). This is believed to be due to the absence of tracheal rigidity, allowing it to be compressed (2).

However, if symptomatic, patients mostly do not present in childhood, but in early adulthood or even older ages. It is believed that symptoms are most often the result of increased oesophageal rigidity, early atherosclerotic changes of the anomalous vessels, dissections or aneurysmal dilatation with compression of surrounding structures (especially in the presence of a Kommerell's diverticulum) (2,4). These alterations cause compression on the surrounding mediastinal structures like the oesophagus, giving rise to mechanical dysphagia, which is called dysphagia lusoria as we have described before.

*Diagnosis of dysphagia lusoria*

Upper endoscopy is usually normal, but occasionally a pulsating extrinsic compression of the oesophagus is seen. Oesophageal manometry frequently reveals unspecific findings (2).

The best method to diagnose dysphagia lusoria is a barium oesophagogram followed by a non-invasive imaging of the chest (2). A barium oesophagogram will show an extrinsic compression of the vascular anomaly. Both CT and MRI are excellent methods for evaluating the mediastinum for vascular anomalies that can cause extrinsic oesophageal compression (8). Both studies have the extra advantage of visualising other possible intra-thoracic pathologies.

*Treatment*

Management of dysphagia lusiora is dependent upon the severity of symptoms. If a patient develops complaints in the absence of a dissection or an aneurysm of the diverticulum, the preferred treatment is a symptomatic one (lifestyle and dietary modification) with close follow-up of the patient (2).

If symptoms persist despite conservative management, surgery can be taken into account. In the presence of a vascular ring, an aneurysm of Kommerell's diverticulum or dissecting thoracic aneurysm surgical or endovascular treatment is mandatory (4,9-11).

## Conclusion

This case report shows the possibility of late manifestations of congenital anomalies. The origin of the complaints in our patient are age-related atherosclerotic changes and dilation of the aorta, giving rise to compression of the oesophagus, which is an explanation for the complaints of dysphagia.

Surgical or endovascular treatment in the absence of an aneurysm of Kommerell's diverticulum or a dissecting aneurysm carry more risks than benefit. This validates the symptomatic treatment in this case report.

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