


## Wheezing and dyspnoea caused by aberrant left innominate artery

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### Keywords

Aberrant left innominate artery, bronchial asthma, right aortic arch, tracheoesophageal compression.

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### Abstract

We present a rare case of a branching anomaly of the aortic arch that resulted in wheezing and dyspnoea. The patient was a 60-year-old male with severe wheezing from babyhood, originally diagnosed with severe bronchial asthma. On auscultation, the inspiratory and expiratory wheezes appeared when the patient leaned forward. He also had difficulty in swallowing solid mass. Tests for airway reversibility and hyperresponsiveness were negative, and asthma treatment was ineffective. He had a right aortic arch. A barium oesophagogram and endoscopic examination indicated narrowing of the oesophagus from behind. Three-dimensional reconstruction of enhanced chest CT images indicated a right aortic arch and an aberrant enlarged left innominate artery, which compressed and narrowed the oesophagus and trachea from behind. Although the patient had been diagnosed with intractable bronchial asthma, his symptoms were more likely caused by this mechanical narrowing as wheezing and dyspnoea disappeared completely after total aortic arch replacement operation.

### Introduction

The aortic arch and associated great arteries originate from six paired embryonic pharyngeal arches, and the final configuration of vascular structure is formed by fusion and regression of these arches. A rare malformation of this system, known as a vascular ring, encircles and compresses the trachea and oesophagus completely or incompletely, causing symptoms to some degree [1].

This report describes a very rare case of a vascular ring with right aortic arch and left innominate artery causing wheezing, dyspnoea, and dysphagia.

### Case Report

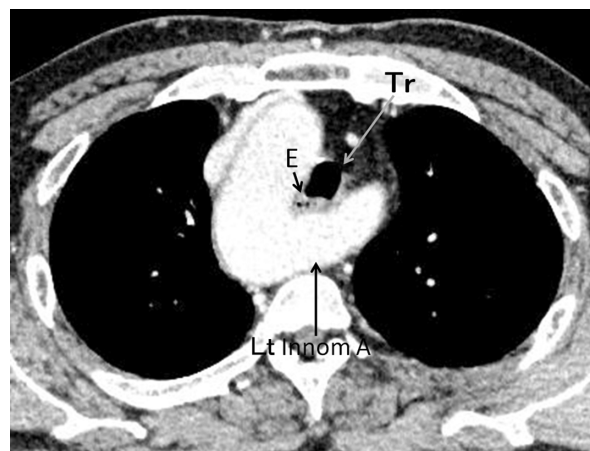
A 60-year-old male was referred to our clinic for preoperative evaluation of bronchial asthma for a cervical spine operation in prone position. Auscultation did not reveal any adventitious sounds in the chest; however, inspiratory and expiratory wheezes were noted while leaning forward

in the sitting position. He could not lie in prone position because of strong dyspnoea. He was a current smoker with a smoking history of 31 pack-years and a history of hypertension and hyperlipidaemia. His medical history indicated that he suffered from severe wheezing from babyhood, which persisted subsequently, especially when crying. He was diagnosed with severe bronchial asthma by his family physician. Despite treatment for asthma, his symptoms persisted. With age, his wheezing became less severe; however, his ability to participate in physical exercise was limited due to increased wheezing and dyspnoea during exertion. Although slight wheezing and dyspnoea occasionally occurred during adolescence, he managed to lead a normal daily life, work in a factory, marry, and have a son. In recent years, however, his symptoms worsened to the point where he became dyspnoeic, even at rest, when suffering from an upper respiratory tract infection. He also experienced dysphagia and heartburn, especially when swallowing solid foods or oral medications. He was diagnosed with a right aortic arch following a regular chest X-

ray check-up. Spirometry indicated that the percentage of predicted vital capacity was 86%, and the ratio of expiratory volume in one second versus forced vital capacity was 72.8%. The bronchodilator test using salbutamol and the methacholine challenge test both yielded negative results, and treatment using oral montelukast and increasing dose of budesonide/formoterol (Symbicort®, AstraZeneca plc, UK) for 1 month was ineffective. These results indicated that the diagnosis of bronchial asthma was an unsuitable explanation for his wheezing and dyspnoea.

To investigate his dysphagia, contrast oesophagography and endoscopy were performed, which indicated that the oesophageal narrowing was caused by a posterior structure (Fig. 1A,B,C).

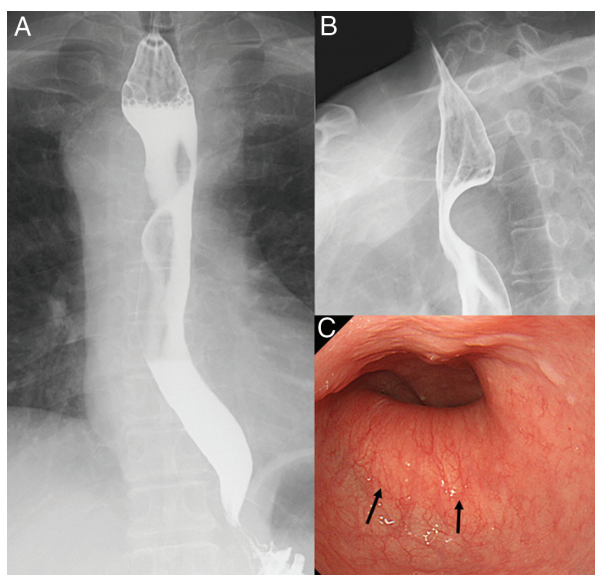
Contrast chest CT revealed a right aortic arch and a thickened left innominate artery encircling the oesophagus and trachea from behind (Fig. 2). A three-dimensional image by volume rendering was constructed (Fig. 3). The ascending aorta (Asc A) arose from the left ventricle (LV), deviated towards the right, and then ran upward to form the right aortic arch. The first branch was the right common carotid artery (Rt CCA), and the vertebral and subclavian arteries (Rt VA and Rt SCA, respectively) branched consecutively from the arch (Fig. 3A). The right aortic arch terminated with the branching of the enlarged left innominate artery (Lt Innom A) and connected with the bulging descending aorta (Des A), as shown in Figure 3B. The left



**Figure 2.** Chest CT with contrast enhancement. Tr: trachea, E: oesophagus, and Lt Innom A: left innominate artery.

innominate artery, which was the remnant of the left aortic arch, encircled the oesophagus and trachea from behind and ran forward parallel to them before diverging into two branches, the left subclavian and the common carotid arteries (Lt SCA and Lt CCA, respectively). There was no ligamentum arteriosum. The oesophagus and trachea were compressed from behind by the enlarged left innominate artery and descending aorta, and significant localized kinking of the oesophagus and trachea was observed. An aneurysm-like bulge was detected at the beginning of the descending aorta, which descended on the right side of the spine until it deviated towards the left side of the spine at the aortic hiatus of the diaphragm.

Total arch replacement operation was performed to relieve the compression of the trachea and oesophagus. Wheezing and dyspnoea completely disappeared after the operation, although slight dysphagia occasionally occurs.

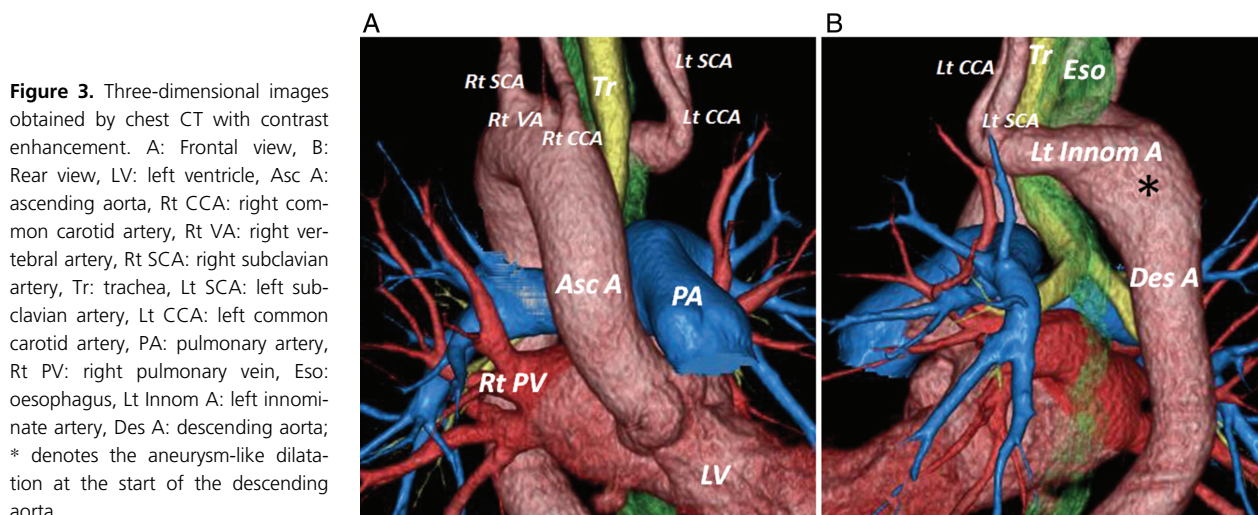


**Figure 1.** Oesophagogram (A: frontal view, B: lateral view) acquired during barium swallow. Depression of the oesophagus and stagnant contrast media are observed. Right aortic arch is also observed. C: Oesophageal endoscopic view. The lumen of the oesophagus is covered by normal epithelium and narrowed by a posterior structure (arrows).

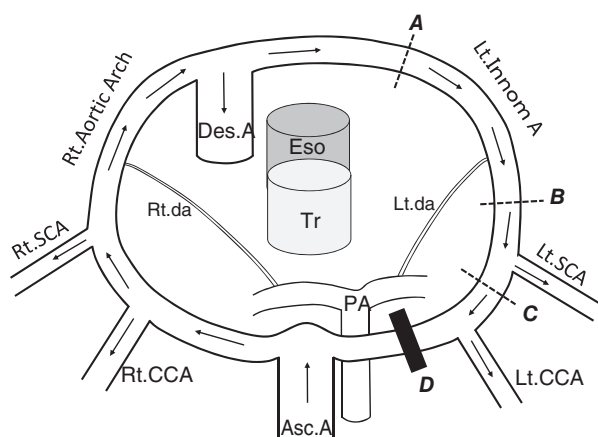
## Discussion

In the case of the right aortic arch, there are four variations for interruption of the left aortic arch and the remnant of the left aortic arch, designated as the left innominate artery [2]. Figure 4 illustrates the major vessels of the vascular ring with a right aortic arch and a right-sided descending aorta. Anomalous interruption of the left aortic arch can occur in any of the four positions (A–D). If an interruption occurs at A or B, the remnant of the left aortic arch remains as Kommerell’s diverticulum (the most common anomaly) [3], whereas the interruption at D, as in this case, is especially rare and has been reported only in isolated cases [4].

In this patient, the symptoms of wheezing and dyspnoea improved during his adolescence, like bronchial asthma would. This was probably due to maturation of the tracheal wall, especially the tracheal cartilage [5]. In recent



**Figure 3.** Three-dimensional images obtained by chest CT with contrast enhancement. A: Frontal view, B: Rear view, LV: left ventricle, Asc A: ascending aorta, Rt CCA: right common carotid artery, Rt VA: right vertebral artery, Rt SCA: right subclavian artery, Tr: trachea, Lt SCA: left subclavian artery, Lt CCA: left common carotid artery, PA: pulmonary artery, Rt PV: right pulmonary vein, Eso: oesophagus, Lt Innom A: left innominate artery, Des A: descending aorta; \* denotes the aneurysm-like dilatation at the start of the descending aorta.



**Figure 4.** Schematic illustration of the right aortic arch and potential sites of interruption (A–D). In the present case, the interruption was at D. Rt.da, and Lt.da denotes the right and left ductus arteriosus, respectively.

years, however, his symptoms worsened, probably as a result of enlargement of left innominate artery and the aorta and the stiffening of the arterial wall by atherosclerosis as he had hypertension and hyperlipidaemia. These structural changes may have played a significant role in the kinking of the trachea and oesophagus as symptoms disappeared after the total aortic replacement operation.

This patient had been misdiagnosed and treated for asthma based only on clinical symptoms. In the case of intractable asthma, a systematic diagnostic assessment

should be warranted in order to exclude alternative diagnosis, such as a vascular ring.

#### Disclosure Statements

No conflict of interest declared.

Appropriate written informed consent was obtained for the publication of this case report and accompanying images.

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