# VASCULAR RING FORMED BY RIGHT AORTIC ARCH WITH ABERRANT LEFT SUBCLAVIAN ARTERY AND LEFT LIGAMENTUM ARTERIOSUM*

## A Rare Cause of Respiratory Distress in Newborn Infants

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**SUMMARY: Ceviz N, Özer S, İkizler C. (Cardiology Unit, Department of Pediatrics, Hacettepe University Faculty of Medicine, Ankara, Turkey). Vascular ring formed by right aortic arch with aberrant left subclavian artery and left ligamentum arteriosum: a rare cause of respiratory distress in newborn infants. Turk J Pediatr 1999; 41: 403-407.**

**Vascular ring, in which the trachea and esophagus are completely surrounded by vascular structures, is one of the causes of respiratory distress in children. Right aortic arch with aberrant left subclavian artery is a common aortic arch anomaly; however, respiratory distress due to vascular ring is seldom associated with this anomaly. We report herein a newborn infant treated surgically because of severe respiratory distress caused by vascular ring formed by right aortic arch with aberrant left subclavian artery and left ligamentum arteriosum. As laryngomalacia was first thought to be the reason for respiratory distress, we suggest that patients with respiratory distress diagnosed with laryngomalacia be evaluated for possible vascular ring.** *Key words: respiratory distress, right aortic arch, vascular ring, laryngomalacia.*

The causes of respiratory distress in children are diverse. Some of them, like vascular rings, may be silent. Several aortic arch anomalies can form a vascular ring and, when sufficient compression of the trachea and/or esophagus exists, symptoms are present[1-3]. When the compression is mild, however, there may be no symptoms[4]. Aberrant subclavian arteries, either right or left, are the most common aortic arch anomalies. However, most of the cases do not have respiratory symptoms; they are rarely associated with respiratory distress.

In this paper, a newborn infant who had been previously diagnosed with laryngomalacia was later found to have a vascular ring. The case is reported to emphasize that patients with respiratory distress and the diagnosis of laryngomalacia must be evaluated for vascular ring. Our patient had right aortic arch with abberrant left subclavian artery and left ligamentum arteriosum, which is an uncommon form of vascular ring.

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## Case Report

A five-day-old newborn infant was admitted to the Neonatology Unit with severe respiratory distress. There was no physical finding related to the cardiovascular system. Chest x-ray showed mild cardiomegaly. Echocardiographic study did not reveal any congenital cardiac abnormalities. Laryngoscopic and fluoroscopic examination of the trachea and larynx suggested laryngomalacia. Barium esophagography revealed a posterior indentation at the first 1/3 proximal part of the esophagus (Fig. 1). Cardiac catheterization was performed. Aortic angiography showed a right aortic arcus and an aberrant retroesophageal left subclavian artery (Fig. 2). Ductus arteriosus was not patent. Since a vascular ring most likely completed by aberrant subclavian artery and ligamentum arteriosum could not be excluded, the patient underwent surgery. The left subclavian artery was the last artery leaving the aortic arch. Left ligamentum arteriosum extending between the left subclavian artery and left pulmonary artery was completing the ring and compressing the trachea. Ligamentum arteriosum was excised and the trachea was released. Although the patient was previously dependent on a ventilator, by

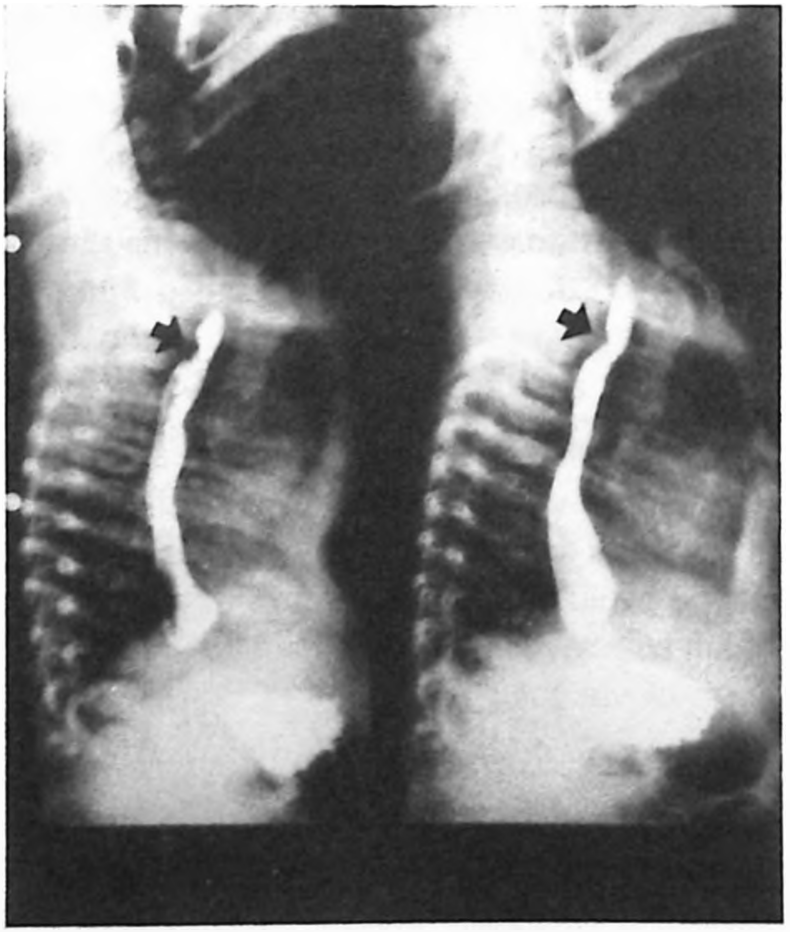
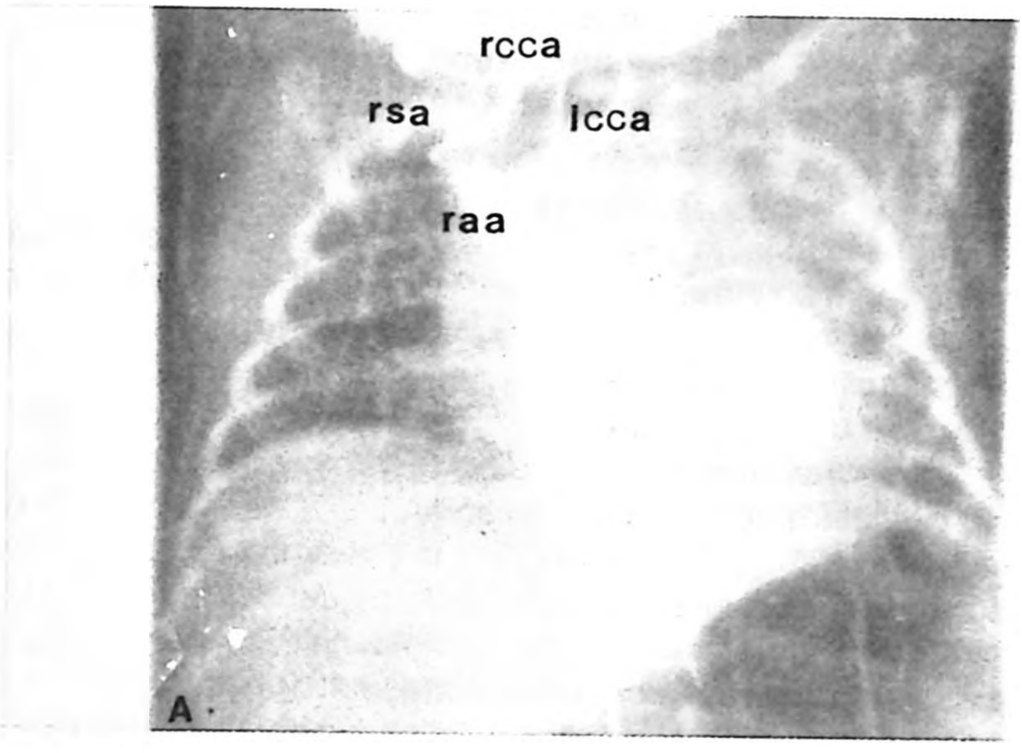
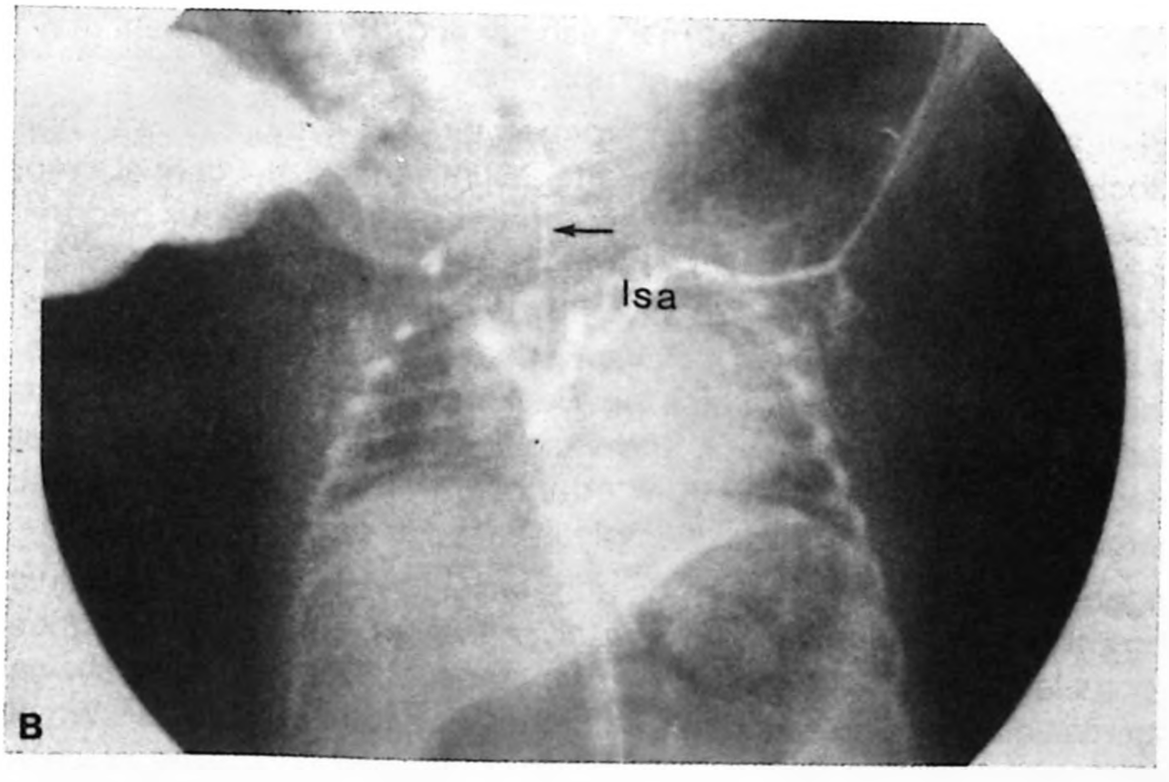


Fig. 1: Barium esophagography depicting the posterior indentation (arrow) at the proximal esophagus.




(a)




(b)

Fig. 2: Aortic angiography at postero anterior projection shows a) right aortic arch (raa), right subclavian artery (rsa), right common carotid artery (rcca), and left common carotid artery (lcca), and b) aberrant retroesophageal left subclavian artery (lsa) stemming from the descending aorta. The arrow indicates the feeding tube placed into the esophagus.

the 48th hour of operation there was no need for artificial ventilation. In the postoperative second week respiratory distress disappeared but pulmonary infection developed; the patient was discharged by the family during treatment.

## Discussion

Vascular ring comprises one to two percent of congenital heart diseases. Although numerous variations from normal aortic arch development have been reported, only a few distinct patterns can produce extrinsic tracheal obstructions. The most likely types that compromise the trachea and esophagus are a) right aortic arch c) anomalous innominate or left carotid artery, d) aberrant right subclavian artery, and e) pulmonary artery sling[5]. The most common aortic arch anomaly causing vascular ring is double aortic arch[1,3]. Cases with right aortic arch are divided into five subgroups according to the branching pattern of the vessels: a) circumflex retroesophageal aortic arch, b) cervical right aortic arch, c) right aortic arch with abnormal origin of left subclavian artery and patent ductus arteriosus, d) right aortic arch with isolated left subclavian artery, and e) right aortic arch with absence of unilateral pulmonary artery. Although aberrant left subclavian artery is the most frequent vascular anomaly seen with right aortic arch, these cases are commonly asymptomatic[3]. Sometimes, as in our patient, a ductus arteriosus or a ligamentum arteriosum may be associated with aberrant left subclavian artery, and may form a vascular ring. Thus, the patient may have symptoms of tracheo esophageal compression.

Reported prevalence of vascular ring formed by right aortic arch with aberrant left subclavian artery and ligamentum arteriosum varies. Lincoln et al.[6] reported 29 patients with vascular anomalies compressing the esophagus and trachea, but none of them had a vascular ring as in our case. Among the 19 children with five types of vascular ring reported by Wychulis et al.[7], only one had right aortic arch with aberrant left subclavian artery and left ductus arteriosus. This patient had respiratory symptoms. In Backer et al's.[8] report, 65 percent of 52 patients with right aortic arch and vascular ring had a retroesophageal left subclavian artery and ligamentum arteriosum. These patients formed the 29 percent of the patients with complete vascular ring.

Laryngomalacia is the most common congenital laryngeal anomaly of unknown etiology that causes inspiratory stridor. Although the clinical diagnosis of laryngomalacia can be confirmed by laryngoscopy, it has been shown that laryngomalacia may be associated with other respiratory disorders[9]. Wu et al.[10] reported seven children with variable types of vascular anomalies, of whom five were previously diagnosed as having laryngomalacia. In Nussbaum et al's study[9], one percent of children diagnosed with laryngomalacia also had vascular ring. Our patient was also first diagnosed as having laryngomalacia. However, after the barium esophagography and angiography, the structural anomaly that was the true reason for respiratory distress was revealed.

In newborn infants with severe respiratory distress, vascular ring must be considered in the differential diagnosis, even if there is a previous diagnosis of laryngomalacia. In these patients, if one detects a right aortic arch with aberrant left subclavian artery, the presence of a vascular ring completed by left ligamentum arteriosum must be considered. These patients may be successfully treated with surgery.

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