

CASE REPORT

Dyspnoea persisting after surgery for a vascular ring

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Dyspnoea persisting after surgery for a vascular ring. A.O. Bron, E.A.M. Mensen, J.H. Dijkman, H.A. Huysmans, R. Aalbers. ©ERS Journals 1994.

ABSTRACT: We present the case of a 25 year old woman who, 3 yrs after resection of a vascular ring, had persisting complaints of episodic dyspnoea. This was caused by compression of the distal trachea and ostium of the right main bronchus by the descending part of the remaining right-sided aorta. Probable mechanisms are discussed.

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Complaints caused by a "vascular ring" are regarded as resulting from entrapment of the trachea and oesophagus [1, 2]. Decompression by resecting part of this ring is, therefore, the recommended treatment, and should give adequate relief [3, 4]. However, we present a case where symptoms persisted 3 yrs after this procedure.

Case report

The patient was a 25 year old female schoolteacher. From the first months of life, difficulty on swallowing, an intermittent stridor and episodes of breathlessness, occurring mainly at meals and when the patient suffered from respiratory infections, were noticed. There was no clear connection between the complaints and body position. At the age of 22 yrs, a vascular ring, consisting of a double aortic arch, was diagnosed. Surgery followed; the left aortic arch and the ligamentum arteriosum were resected. Afterwards, the stridor and dysphagia disappeared.

However, the episodes of dyspnoea, still mainly associated with infections, continued. Because of intermittent obstruction on ventilatory function testing, a history of hay-fever and signs of atopy, a tentative diagnosis of asthma was made, although there was no strong connection between the complaints and allergic provocation. Also, bronchial hyperreactivity could not be convincingly demonstrated. Bronchodilators and inhaled steroids were prescribed, and as the severity and frequency of complaints increased, oral prednisone was added. Even then, the episodes of dyspnoea persisted. The patient was referred to the Netherlands Asthma Centre, Davos, Switzerland.

On admission, she was not dyspnoeic, there was no stridor, and on auscultation normal breath sounds were

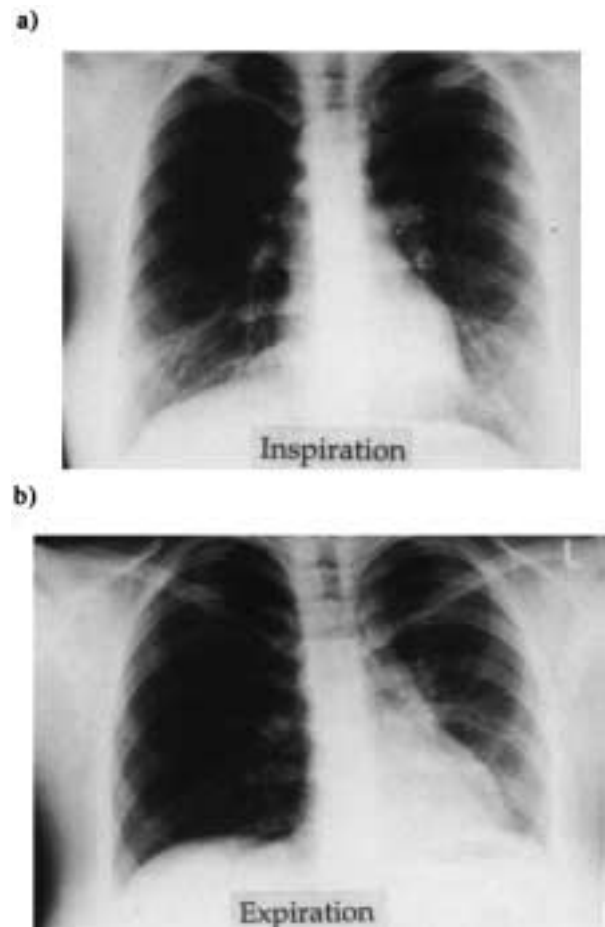


Fig. 1. - a) Inspiratory and b) expiratory roentgenograms of the chest. On expiration, the right hemi-diaphragm does not move upward and the mediastinum shifts to the left, indicating air-trapping in the right lung. Note the right-sided aorta.

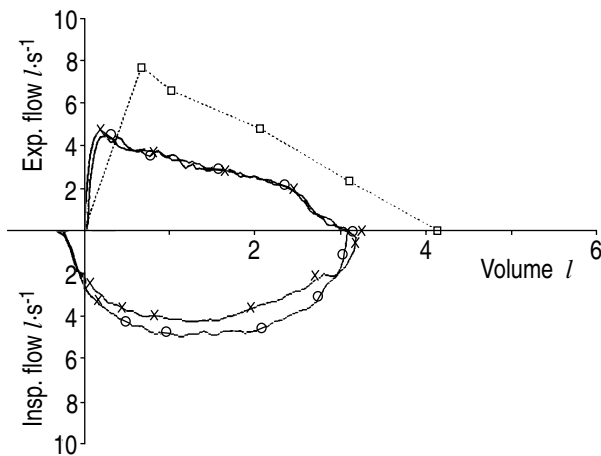


Fig. 2. — Maximal flow-volume curves with the expiratory part showing the typical findings associated with intrathoracic obstruction of the large airways.: predicted values; \circ : prebronchodilator values; \times : postbronchodilator values. Insp.: inspiratory; Exp.: expiratory.

heard on both sides of the chest. Laboratory results were unremarkable, apart from a raised immunoglobulin E (IgE) level $430 \text{ kU}\cdot\text{l}^{-1}$, (reference value $<150 \text{ kU}\cdot\text{l}^{-1}$). Radioallergosorbent test (RAST) showed a class 3 reaction for orchard grass.

A roentgenogram displayed no abnormalities, apart from the right-sided aorta. On ventilatory function testing, no significant obstruction was found; however, the mid-expiratory part of the flow-volume curve had a slightly "flattened" appearance.

Several weeks after admission, the patient reported progressive dyspnoea in association with a respiratory infection. A wheeze over the distal trachea and almost absent breathing sounds over the right lung, especially noticeable on expiration, were now evident. The roentgenogram revealed air-trapping on the right side (fig. 1). Ventilatory function testing showed much more distinctive "flattening" of the expiratory part of the flow-volume curve, suggestive of intrathoracic obstruction of the large airways (fig. 2) [5]. Also, forced expiratory

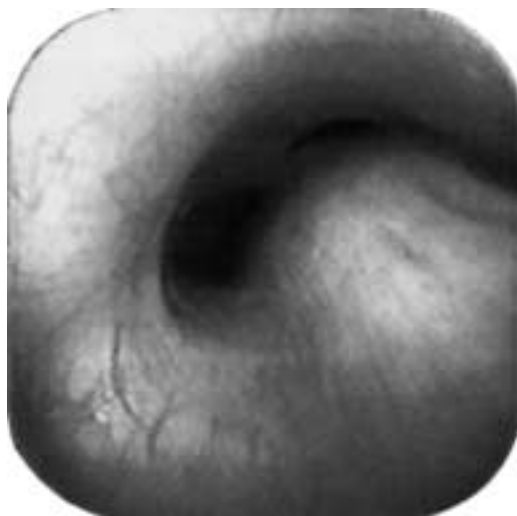


Fig. 3. — Endobronchial view of the distal trachea showing compression, especially of the right main bronchus, from the right dorsal side.



Fig. 4. — Computed tomographic (CT) scan of the thorax showing the distal trachea compressed by the descending right-sided aorta (arrow). Note that the aorta at this point crosses the spinal column, which prevents posterior movement of this vessel.

volume in one second (FEV_1) and forced vital capacity (FVC) were decreased: 65 and 73% of predicted, respectively [6]. There was no reversibility after inhalation of salbutamol.

The patient was treated with high dose intravenous prednisolone. Subsequently, the clinical situation improved. Bronchoscopy was performed; expiratory compression of the distal trachea and particularly the ostium of the right main bronchus was seen. This was caused by a pulsatile indentation from the right dorsal side (fig. 3). Computed tomography (CT) showed this to be the descending arch of the right-sided aorta (fig. 4).

Treatment was surgical. The aortic arch was partially resected and replaced by an elongated graft to prevent renewed compression. After this operation, the patient became symptom-free.

Discussion

Vascular rings are a rare but well-known cause of respiratory complaints. A double aortic arch, with both systems patent, is the most common variety [3]. It is stated that constriction of the airways and the oesophagus is caused by the tightness of the vascular ring [2]. Usually, this condition is diagnosed in the first year of life, but later detection has been reported [2, 7, 8]. As in our patient, complaints are sometimes intermittent and occur only under special circumstances [2, 7].

If significant symptoms exist, surgical treatment is recommended [3]. It is said that this procedure is highly successful and that relief of the respiratory problems is predictable [4]. Although the left aortic arch and the ligamentum arteriosum were resected 3 yrs previously, our patient still had respiratory complaints. This was caused by continuing compression of the airways by the descending arch of the (right-sided) aorta. To our knowledge, this has not been described previously.

We now discuss the probable mechanisms. Firstly, the specific anatomical circumstances created by a right-sided aorta must be considered. A right-sided aorta may descend on the left or the right side [3]. If descending

on the left side, this vessel must cross the spinal column. At this point it cannot move backwards and must compress the airways, which are bound in position by the pulmonary artery, the superior vena cava, the vena azygos and the ascending arch of the aorta itself. In our case, CT and bronchoscopy clearly show that compression takes place at this point (figs. 3 and 4).

In addition, when a vascular ring is still intact the airways are compressed by the right-sided aorta at this point, as can be seen on illustrations provided in several case reports, one of which was published in this Journal [2, 7, 8]. Also, WRIGHT and ALEXANDER [9] described a case where a tortuous left-sided aorta, in combination with kyphoscoliosis and in the absence of a vascular ring, caused severe respiratory problems by the same mechanism.

Other factors are probably contributory. In our patient, inflammatory oedema caused by infection or an asthmatic reaction, seems to have acted as a "triggering" factor, presumably by critically narrowing the lumen at the point of compression.

Once significant obstruction exists, raised intrathoracic pressures may further complicate the situation. Airway collapse would of course most easily occur at the point where the aorta, by compressing the airways, has possibly already induced malacia. Indeed in our patient, slight tracheomalacia was found on reoperation. However, it was judged that this could not have caused significant obstruction. Furthermore, such a malacia is reported to cause respiratory problems mainly in the first weeks, or at most months, after operation [3].

No signs of mediastinal fibrosis were seen in our patient at the second operation, thus excluding renewed constriction by this mechanism.

In conclusion, we would like to offer the following suggestions. Every patient with a right-sided aorta, particularly when this situation results from surgical correction of a vascular ring, should be carefully followed up. In this respect, ventilatory function testing and inspection of the flow-volume curve are especially important. When necessary, bronchoscopy and/or various imaging techniques should be applied as well.

Especially in cases where the descending aorta crosses over the spinal column, compression of the airways should

be excluded before alternative hypotheses to explain respiratory symptoms are entertained. During surgery for a vascular ring, the possibility of compression of the airways by the right-sided aorta should be appreciated and additional measures taken whenever appropriate. However, in practice this might be difficult as the conventional surgical approach is from the left side [3].

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References

1. Lincoln JCR, Deverall PB, Stark J, Aberdeen E, Waterson DJ. Vascular anomalies compressing the oesophagus and trachea. *Thorax* 1969; 24: 295-306.
2. Bevelacqua F, Schicchi JS, Haas F, Axen K, Levin N. Aortic arch anomaly presenting as exercise-induced asthma. *Am Rev Respir Dis* 1989; 140: 805-808.
3. Wychulis AR, Kincaid OW, Weidman WH, Danielson GK. Congenital vascular ring: surgical considerations and results of operation. *Mayo Clin Proc* 1971; 46: 182-188.
4. Salzberg AM, Krummel ThM. Congenital malformations of the lower respiratory tract. In: Chernick V, Kendig EL, eds. *Disorders of the Respiratory Tract in Children*. 5th edn. Philadelphia, WB Saunders Co. 1990; p. 240.
5. Pedersen OF, Ingram RH Jr. Configuration of maximum expiratory flow-volume curve: model experiments with physiological implications. *J Appl Physiol* 1985; 58: 1305-1313.
6. Quanjer PhH, ed. Standardised lung function testing. *Bull Eur Physiopathol Respir* 1983; 19 (Suppl. 5): 1-95.
7. Bose S, Hurst TS, Cockcroft DW. Right-sided aortic arch presenting as refractory intraoperative and postoperative wheezing. *Chest* 1991; 99(5): 1308-1310.
8. Dahlen IB, Hillerdal GN, Wegenius GA, Henze AC. Breathlessness in a teenager. *Eur Respir J* 1993; 6: 145-147.
9. Wright PM, Alexander JP. Acute airway obstruction, hypertension and kyphoscoliosis. *Anaesthesia* 1991; 46(2): 119-121.