

Airway Narrowness Affects the Severity and Onset of Respiratory Symptoms in Patients with Vascular Ring

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Introduction Vascular ring is an uncommon congenital aortic arch anomaly where trachea and esophagus are compressed by the surrounding vascular structures and cause progressive respiratory and/or esophageal symptoms during infancy and childhood. Variable degree of respiratory symptoms has been attributed to the tightness of the ring, but the relationship between the two has not been established. We tested whether the degree of tracheal obstruction correlates with the severity of respiratory symptoms in patients with vascular ring.

Methods We retrospectively reviewed 61 patients with isolated vascular ring who underwent surgical repair and studied their clinical presentation and the anatomical size of trachea at the time of diagnosis. The respiratory symptoms were classified as I) no symptoms ($n = 19$), II) mild symptoms (cough, snoring, loud breathing, or stridor; $n = 21$), III) moderate symptoms with respiratory functional abnormalities (increased work of breathing or recurrent respiratory infection; $n = 17$), and IV) life threatening conditions (cyanosis, apnea, or cardiopulmonary arrest; $n = 4$). The airway diameter was measured by either CT or MRI. Tracheal narrowing was assessed by the ratio of the narrowest tracheal diameter (TS) to the narrowest diameter at the thoracic inlet (TI).

Results The onset of symptoms was earlier (median of 11.5 weeks) in IV compared to 9 months and 3 years in group III and II, respectively (both $p < 0.05$). Among symptomatic groups, there was weak positive correlation between the severity of symptoms and the degree of tracheal stenosis. Post-operative hospital days tended to be longer in patients with severe symptoms (II: 3.3 ± 1.2 days, III: 6.3 ± 5.8 days, and IV: 8.0 ± 2.0 days). Group I showed the widest variation of the all groups.

Conclusions Although the degree of tracheal narrowing did not correlate with either onset or severity of symptoms, patients with severe respiratory symptoms tended to become symptomatic earlier. There may be other factors contributing to the severity of respiratory symptoms.

