Vascular Tracheobronchial Compression Syndrome in Adults: A Review

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(Received July 20, 2011; Accepted August 11, 2011)

Key words: Vascular tracheobronchial compression syndrome, vascular ring,aortic arch anomalies, aortic arch aneurysm, flow-volume curve

Respiratory symptoms caused by tracheobronchial compression by the aorta is called vascular tracheobronchial compression syndrome, and rarely symptoms first appear during adulthood [1]. Adult patients are sometimes followed for many years with a diagnosis of bronchial asthma, because wheezes and dyspnea are also the major symptoms of tracheobronchial compression. In some cases, the diagnosis was made after the airway has been severely remodeled and their lung function had deteriorated, thus cannot be cured surgically [2]. Therefore it is of clinical importance to understand the pathophysiology of tracheobronchial vascular compression syndrome and to make an early diagnosis of it. Although there is one excellent review describing vascular rings of the thoracic aorta in adults [3], it included esophageal symptoms as well as respiratory ones. Also it has now been over a decade since then. Here, we will review this syndrome highlighting respiratory symptoms.

Several cases of vascular tracheobronchial compression syndrome in children due to aortic arch anomalies have been reported [4, 5]. Keith *et al*. [6] claimed that it accounts for 1.2 percent of congenital heart disease in children. However, cases primarily found in adults are much rarer. Sebening *et al*. [1] reported that there were only 2 adults among 22 cases of tracheobronchial compression by the aorta that they operated on who were 34 and 46 years old. Only fragmentary information exists concerning prevalence of vascular tracheobronchial compression syndrome in adults, but it is considered to be very rare.

Vascular tracheobronchial compression syndrome in adults is grouped into congenital and acquired types. Congenital type may be anomalies that are primarily asymptomatic and found incidentally during adulthood, or may cause symptoms only when the aorta becomes rigid due to atherosclerosis [7, 8]. Most cases of the acquired type are due to aortic aneurism, but kinked aorta can also produce compression [2, 9].

CONGENITAL TYPE

To better understand the anatomy of vascular tracheobronchial compression, development of large arteries in embryonic terms should be briefly described. During embryogenesis, mediastinal large arteries form six symmetrical paired aortic arch vessels and the paired dorsal aorta (Fig. 1). Normally the first and second aortic arches dissolve. The third arch form right and left common carotid arteries and part of internal carotid arteries. The left fourth arch persists as aortic transverse arch, and the right fourth aortic arch leads to the right arm via dorsal aorta and the seventh intersegmental artery. The fifth arches disappear in early stage. The right sixth arch loses contact with the dorsal artery and becomes the right pulmonary artery, while the left sixth aortic arch persists as the ductus arteriosus. If the communication between the right sixth arch and the dorsal artery persists, these vessels form a vascular ring that encircles esophagus and trachea, i.e., bilateral ductus arteriosus.

Double aortic arch is a persisting communication between the fourth arch and the right dorsal artery after embryogenesis, and this produces a vascular ring (Fig. 2A).

Right aortic arch is persisting right fourth arch instead of left fourth arch which is supposed to form the aorta. Pulmonary artery is formed normally but the aorta presents with mirror image of the normal. It doesn't cause compression itself but if the ductus arteriosus connects to the left pulmonary artery, the trachea and esophagus are encircled between the aortic arch and ductus or ligamentum arteriosus (Fig. 2B). Right aortic arch may accompany aberrant left subclavian artery in which left subclavian artery arises from right-sided descending aorta. Ductus arteriosus may originate from the base of normal left subclavian artery, which is more common, or it may arise from the remaining diverticulum at the base of the eighth dorsal artery during embryogenesis (Kommerell's diverticulum) (Fig. 2C).

Aberrant right subclavian artery which arises on the left side of the left-sided aorta and runs posterior to the trachea and the esophagus, also results in a vascular ring (Fig. 2D).

Edwards' classification of aortic anomalies is frequently used which is based on hypothetical double aortic arch system (Table 1). Right aortic arch was further classified by Stewart *et al*. into more detailed

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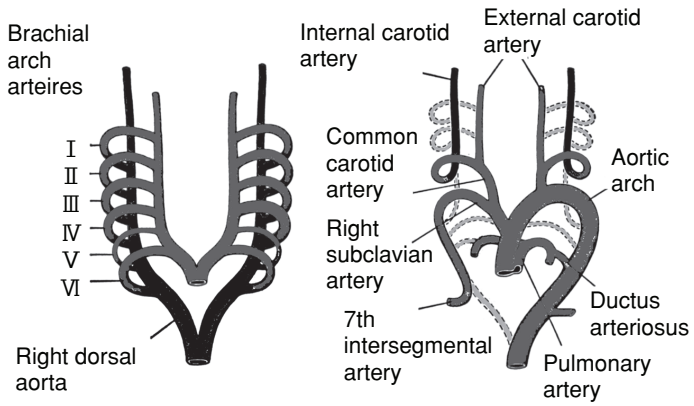

Fig. 1 Anatomy of aortic arch system during embryogenesis

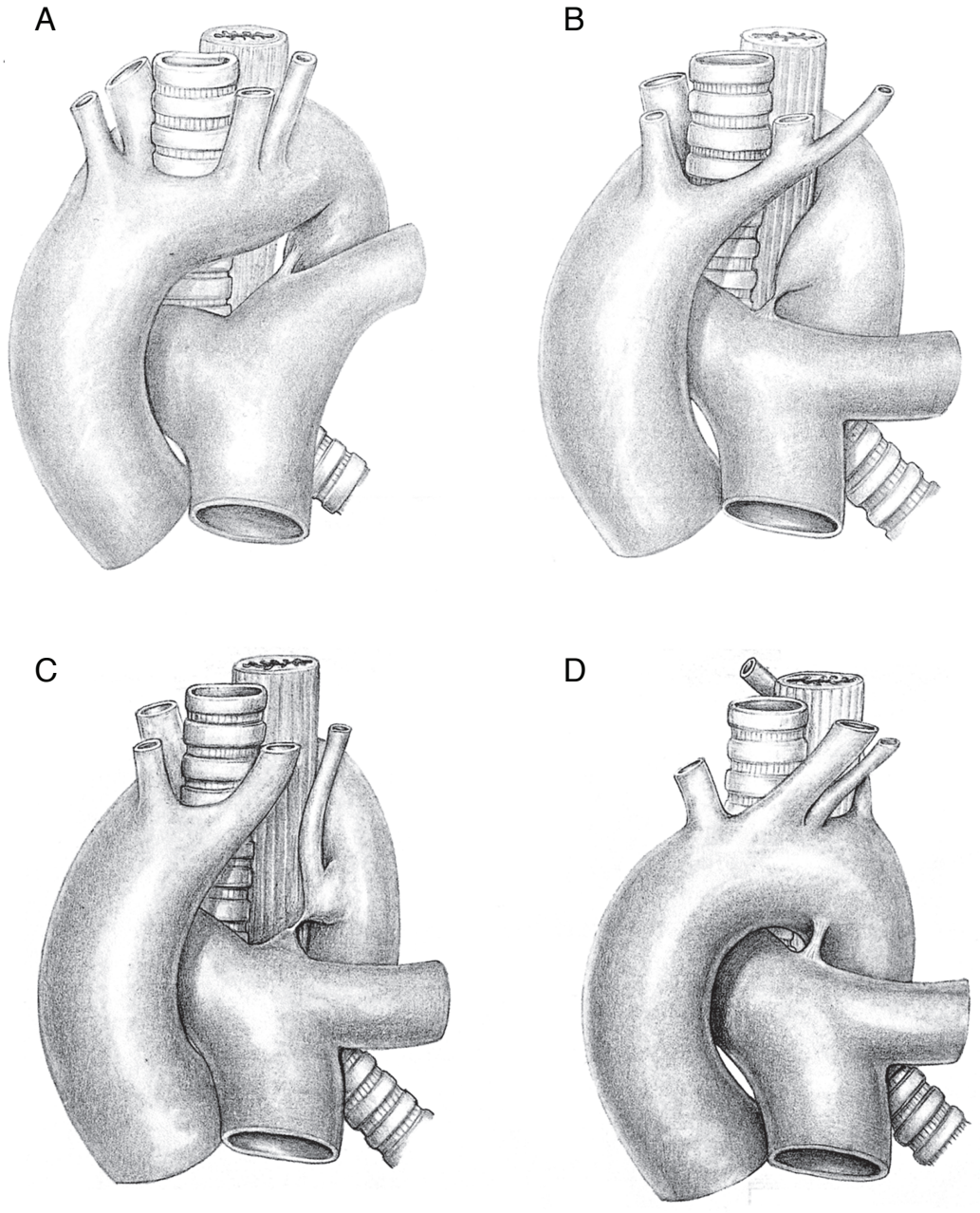

Fig. 2 Types of vascular rings [11]. A, Double aortic arch. B, Right aortic arch (Mirror-image arch). C, Right aortic arch with aberrant left subclavian artery that arises from Kommerell's diverticulum. D, Aberrant right subclavian artery

Table 1 Edward's classification of the aortic arch anomalies

double aortic arch	A: both aortic arch patency
〃	B: one side aortic arch close
left aortic arch	A: normal branch
〃	B: aberrant right subclavian artery
〃	C: isolation of right subclavial artery
right aortic arch	A: mirror image branch
〃	B: aberrant left subclavian artery
〃	C: isolation of left subclavian artery
Others	

Each subgroup is classified in side of ductus arteriosus.
1: left arteriosus
2: right arteriosus
3: bilateral arteriosus

Table 2 Congenital vascular anomalies causing tracheobronchial compression

	Sebening(1)	Ruzmetov(7)	Woods(5)
1. Double aortic arch	7	67	31 (38%)
2. Right aortic arch	5	77	22 (27%)
1 + 2	12 (55%)	144 (78%)	53 (65%)*
3. Pulm. artery sling	3	8	3 (4%)
4. Aberrant rt subclavian artery	-	30	4 (5%)
5. Others	7**	1	22 (26%)***

*Others are excluded in calculation
**status post ligation/transaction of patent ductus arteriosus 2, status post repair of coarction of patent ductus arteriosus 1, abnormally inserted ligamentum arteriosum 1, rt lung aplasia + lt ligamentum arteriosum 1
***Innominter artery compression 20 and aberrant left subclavian artery 2

anatomical types [10].

Table 2 demonstrates classification and prevalence of congenital vascular tracheobronchial compression including both children and adults reported by Sebening, Woods, and Ruzmetove [1, 5, 7]. Most subjects in those reports were children; Only 2 were adults (34 and 46 years old) who had right aortic arch reported by Sebening, all were 12 years old and below by Wood [5], and age ranged from 3 to 30 years by Ruzmetove with median of 5 months old [7]. Approximately fifty to eighty percent of all cases were either double aortic arch or right aortic arch. Wood *et al*. [5] reported that other than compression by the vascular ring, 20 out of 82 had compression by innominate artery. Yasui [11] described that among 160 cases of vascular rings [12] 68% were double aortic arch, 19% were right aortic arch with left ductus arteriosus, and 11% were aberrant right subclavian artery.

Table 3 is a list of reported adult cases of vascular ring in which respiratory symptoms were confirmed. Among 36 cases, 22 were diagnosed right aortic arch, and 10 were diagnosed double aortic arch. This ratio is different from that in children, in which double aortic arch is dominant [5]. Most common symptoms were wheezing, dyspnea, and dysphagia, which are same as cases of children. In adult-onset cases, dyspnea is more common than dysphasia and it may mimic asthma, though dysphagia often gets serious [13, 14]. Double aortic arch typically cause significant tracheal and esophageal symptoms and need surgical correction during childhood, while right aortic arch may escape detection [15]. This may explain why right aortic arch is dominant to double aortic arch in adults.

ACQUIRED TYPE

The most prevalent kind of acquired vascular tracheobronchial compression syndrome is aneurism compressing the tracheobronchial tree, and many cases have been reported [16]. Others reported cases of brachiocephalic artery aneurism [17]. Although most aneurisms result from atherosclerosis, aortic aneurism is often associated with Marfan syndrome in young adults [18]. When an aneurism is present in the ascending aorta, syphilis may be suspected [19]. Airway compression due to aortic aneurism usually presents with wheezing, coughing, hemoptysis, dyspnea and recurrent pneumonia [16]. Kinked aorta due to atherosclerosis or scoliosis can compress the major trachea [2], however there has been only one case reported [9].

DIAGNOSIS

Both congenital and acquired vascular compression syndrome present with wheezing and dyspnea. Mechanisms that cause these symptoms are complex,

Table 3 Cases of adult-onset congenital anomlies that cause tracheobronchial compression with severe respiratory symptoms or required surgical therapy.

No	Anomaly type	age	sex	Respir. sympt.	author	year	Ref.
1	Rt. aortic arch	?	?	yes	Okuda	1986	[30]
2	Double Aortic Arch	50	F	?	Kron	1987	[31]
3	Rt. aortic arch	?	F	yes	Bevelaqua	1989	[32]
4	Double Aortic Arch	78	M	yes	Mori	1992	[33]
5	Double Aortic Arch	67	M	yes	Midulla	1992	[34]
6	Double/rt. aortic arch	24	F	yes	Bron	1994	[21]
7	Rt. aortic arch	32	F	yes	Fadel	1995	[35]
8	Rt. aortic arch	?	?	yes	van Son	1999	[36]
9	Rt. aortic arch	?	?	yes	van Son	1999	[36]
10	Double Aortic Arch	36	M	yes	Grathwohl	1999	[3]
11	Double Aortic Arch	29	F	?	Brockes	2000	[37]
12	Rt. aortic arch	46	F	yes	Sebening	2000	[1]
13	Rt. aortic arch	34	F	yes			
14	Double Aortic Arch	30	F	yes	Stoica	2000	[22]
15	Rt. aortic arch	67	F	yes	Lunde	2002	[38]
16	Rt. aortic arch	76	F	yes	Hardin	2003	[20]
17	Rt. aortic arch	65	F	yes	Greiner	2003	[39]
18	Rt. aortic arch	21	M	yes	Chujo	2004	[40]
19	Rt. aortic arch	42	F	yes	Winn	2004	[23]
20	Double aortic arch	39	M	yes	Kondo	2005	[41]
21	Double aortic arch	49	F	yes	Ohhashi	2005	[42]
22	Rt. aortic arch	44	F	yes	Grillo	2006	[43]
23	Rt. aortic arch	26	F	yes	Grillo	2006	[43]
24	Rt. aortic arch	?	?	yes	Bashar	2006	[44]
25	Rt. aortic arch	?	?	yes			
26	Rt. aortic arch	?	?	yes			
27	Lt. carotid artery anomaly	23	M	yes	Joshi	2006	[45]
28	Rt. aortic arch	47	F	yes	Hickey	2007	[28]
29	Double aortic arch	69	F	yes	Lee	2007	[46]
30	Rt. aortic arch	53	F	yes	Ko	2007	[47]
31	Rt. aortic arch	75	F	yes	Miyamoto	2008	[2]
32	Rt. aortic arch	54	F	yes	Tjang	2008	[48]
33	Double aortic arch	52	M	yes	Choh	2008	[49]
34	Rt. aortic arch	40	F	yes	Sato	2009	[50]
35	unknown	30	?	?	Ruzmetov	2009	[7]
36	Pulmonary artery sling	42	F	yes	LaBelle	2010	[51]

and Hardin *et al* blamed it on both direct compression and secondary weakness of the airway walls [20]. There are dynamic and static mechanisms that compression is caused. During exercise, the diameter of the descending aorta increased by approximately 1 mm or more for every 15 mmHg increases in systolic or diastolic pressure, thus narrowing the airway by 3 to 6 mm in circumference [20]. At the point when patients are aware of respiratory symptoms, pulmonary function may be remarkably impaired already, and it is sometimes hard for these patients to perform spirometry. Flow-volume curve may demonstrate flattening of the expiratory portion (Fig. 3), suggesting variable intrathoracic obstruction [21–23]. Diameter of the aorta increases by 1 mm every decade due to normal aging, besides atherosclerosis and hypertension increase tortuosity of the aorta [20], these elements should be taken into account in elderly-onset patients.

As for radiographic testing, chest X-ray may reveal enlargement of the upper mediastinum. Most cases accompany esophageal compression, so contrast study of the esophagus would also help. The easiest test is chest CT scan, and vascular tracheobronchial compression syndrome can be confirmed with little physical

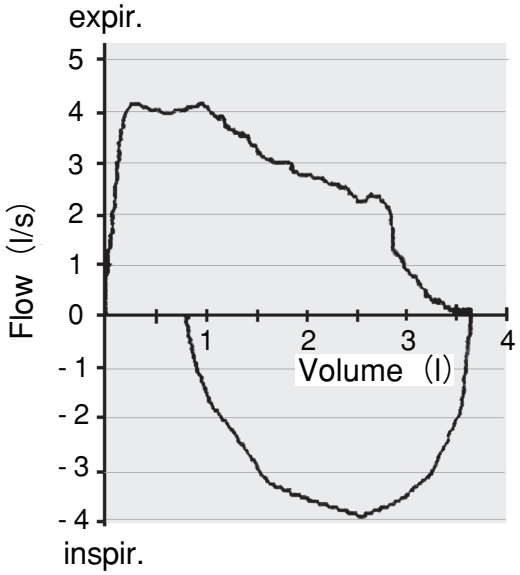

Fig. 3 A, An example of flow-volume curve of tracheal vascular compression syndrome [23]. Note, flattening of the expiratory segment.

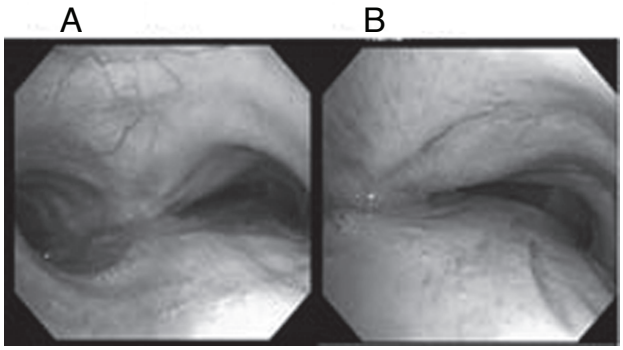

Fig. 4 Right main bronchus obstruction seen in an asymptomatic patient with right aortic arch. A, Tracheal bifurcation. B, Right main bronchus. The upper part of the screen is anterior.

invasion by virtual bronchoscopy, which is three-dimensional reconstruction by CT scan. Actually, bronchoscopy often reveals right aortic arch incidentally in asymptomatic patients (Fig. 4), however, it is speculated that in many cases surgical correction cannot be justified. MRI angiography is also a promising method to detect tracheobronchial compression by large arteries [24].

TREATMENT

As for vascular rings in adults, principle of surgical correction is the same as children. Right aortic arch need transection of ligamentum arteriosum, but only this procedure may not be enough to decompress the trachea. The aorta distal to right common carotid artery may need to be suspended forward in order to release pressure, and some adults need reinforcement of the trachea [1, 17]. Double aortic arch needs division of one of the arches that form the ring. Basically the larger arch is preserved, however, in order to relieve compression, it is desirable that posterior, rather than anterior, arch is transected. Sometimes the remaining arch has to be sutured to the sternum to avoid tracheal compression.

In some cases tracheal intubation was not easy because the trachea had become narrow, pressure inside trachea during mechanical ventilation was too high [19], or some elderly patients cannot be operated on because their pulmonary function had deteriorated [2]. Besides, in a case in which compression was present for a long time, there persisted tracheobrochomalacia after surgical repair and needed enforcement of the trachea [17].

Intra-luminal airway stent is considered to dilate narrowed airway uninvasively. There have been cases in which airway stent was placed in emergency settings for tracheobronchial compression caused by an aneurism [16] and by vascular ring [2], however, there's no consensus on whether airway stenting for benign diseases can be justified [25]. Especially, for narrowed trachea due to vascular compression, the airway will always be exposed to mechanical stimuli therefore the stent needs to be very strong. Also there's no knowing if the tracheal wall can stand mechanical pressure both from inside by the stent and from outside by the aorta [26, 27]. Propensity of granulation and stricture development should also be considered [28, 29].

We emphasize that some adult patients are hard to treat after compression has lasted for many years. So if a patient has right sided aorta and suffers an asthma-like symptoms, this rare condition should be included in the differential diagnosis and you should not hesitate to perform virtual bronchoscopy, so that vascular tracheobronchial compression syndrome can be diagnosed at an early stage.

REFERENCES

1) Sebening Ch, Jakob H, Tochtermann U. *et al*. Vascular tracheobroncial compression syndromes. Thorac Cardiov Surg 48: 164-174, 2000.
2) Miyamaoto Y, Ohashi A, Ishii H, *et al*. A case of severe bronchial stenosis caused by a vascuylar ring in an elderly woman. J Jpn Respir Soc 46: 243-247, 2008.
3) Grathwoul KW, Afifi AY, Dillard TA, *et al*. Vascular rings of the thoracic aorta in adults. Am Surg 65: 1077-1083, 1999.
4) Hasegwa M, Ohta H, Omata M, *et al*. A case of vascular ring. Clini Pediat 49: 2029-2034, 1996.
5) Woods RK, Sharp RJ, Holcomb GW, *et al*. Vascular anomalies and tracheoesophageal compression: a single institution's 25-year experience. Ann Thorac Surg 72: 434-439, 2001.
6) Keith JD *et al*. Heart disease in infancy and childhood. 3rd ed. New York. Macmillian. 1978.
7) Ruzmetov M, Vijay P, Rodefeld MD, *et al*. Follow-up of surgical correction of aortic arch anomalies causing tracheoesophageal compression: a 38-year single institution experience. J Pediat Surg 44: 1328-1332, 2009.
8) Yusa H, Nishiya Y, Murata A. Severe tracheal compression resulting from atherosclerotic aortic arch aneurysm: report of a case. Kyobu Geka 58: 1077-1080. 2005
9) Wright PM, Alexander JP. Acute airway obstruction and kyphoscoliosis. Anesthesia 46: 119-121, 1991.
10) Stewart JR, Kincaid OW, Titus JL. Right aortic arch: plain film diagnosis and significance. Am J Roent, radium Ther, Nuclear Med 97: 377-389, 1966.
11) Yasui H. Vascular Ring. In: Surgery of congenital heart anomaly ed. Sumi H and Masuda M. Medical View, Tokyo, 2003 pp42-43.

12) Haryansky IL, Lozasdi M, Marcsek P, *et al*. Congenital vascular rings: surgical management of 111cases. Eur J Cardiovasc Surg 3: 250–254, 1989.
13) Kouchoukos, N *et al*. Kirklin / Barratt-Boyes Cardiac Surgery, 3rd ed., Churchill Livinston. 2003.
14) Boesler M, De Leval M, Chrispin A, *et al*. Surgical management of vascular ring. Ann Surg 197: 139–146, 1983.
15) Takao A. Clinical and developmental cardiology, 3rd ed. p606. Chugai Igaku-sha, Tokyo 2000.
16) Chen W-C, Tu C-Y, Liang S-J, *et al*. Metalic stents for a patient with severe upper airway compression due to aortic aneurism. Am J Emerg Med 27: 256e1–e4, 2009.
17) Matushita T, Nakamura A, Kawachi H, *et al*. A case report of surgical treatment for aneurysm of the brachiocephalic artery associated with tracheostenosis. J Jpn Assn Thorac Surg 45: 754–758, 1997.
18) Takara I, Uehara M, Higa Y, *et al*. Respiratory management in a patient with severe tracheal stenosis caused by compression from the ascending aortic arch aneurysm. Masui 52: 1079–1082, 2003.
19) Tominaga R, Tanaka J, Wawachi Y, *et al*. Surgical treatment of respiratory insufficiency due to tracheobronchial compression by aneurysm of the ascending aorta and innominate artery. J Cardiovasc Surg 29: 413–417, 1988.
20) Hardin RE, Brevetti GR, Sanusi M, *et al*. Treatment of symptomatic vascular rings in the elderly. Tex Heart Inst J 32: 411–415, 2003.
21) Bron AO, Mensen EAM, Dijkman JH, *et al*. Dyspnoea persisting after surgery for a vascular ring. Eur Respir J 7: 2257–2259, 1994.
22) Stoica SC, Lockwandt U, Coulden R, *et al*. Double aortic arch masquerading as asthma for thirty years. Respiration 63: 92–95, 2002.
23) Winn RA, Chan ED, Langmack EL, *et al*. Dyaphagia, chest pain, and refractory asthma in a 42-year-old woman. Chest 126: 1694–1697, 2004.
24) Teo LLS, Hia CPP. Advanced cardiovascular imaging in congenital heart disease. Internat J Clin Pract. 65: s17–S29, 2011.
25) Madden BP, Loke T-K, Sheth AC. Do expandable metallic airway stents have a role in the management of patients with benign tracheobronchial disease? Ann Thorac Surg 82: 274–278, 2006.
26) Wadsworth SJ, Juniper MC, Benson MK, *et al*. Fatal complication of an expandable metallic bronchial stent. British J Radiol 72: 706–708, 1999.
27) Nourarei SM, Pillay T, Hilton CJ. Emergency management of aorto-bronchial fitula after implanation of a self-expanding bronchial stent. Eur J Cardio-thorac Surg 20: 642–644, 2001.
28) Hickey EJ, Khan A, Anderson D, *et al*. Complete vascular ring presenting in adulthood: an unusual management dilemma. J Thorac Cardiovasc Surg 134: 235–236, 2007.
29) Tazaki G, Kondo T, Kamio K, *et al*. Crescent moon-type tracheobronchomalacia alleviated by placement of stents in trachea and main bronchi. J Jpn Respir Soc 38: 476–479, 2000.
30) Okuda O, Yamamoto S, Kotani I, *et al*. A case of vascular ring of Stewart & Edwards IIIB-1 in an adult. Jpn Circ J 50 (suppl) 208: 12–20, 1986.
31) Kron IL, Mappin G, Nolan SP. Symptomatic double aortic arch causing tracheal and esophageal compression in the adult. Annals Thorac Surg 43: 105–106, 1987.
32) Bevelaqua F, Schicchi JS, Hass F *et al*. Aortic arch anomaly presenting as exercise-induced asthma. Am Rev Respir Dis 140: 805–808, 1989.
33) Mori H, Takeuchi Y, Gomi A, *et al*. An emergency surgical case of double aortic arch with hemoptysis. Nippon Kyobu Geka Gakkai Zasshi 40: 1157–1160, 1992.
34) Midulla PS, Dapunt OE, Sadeghi AM, *et al*. Aortic disection involving a double aortic arch with a right desending aorta. Ann Thorac Surg 58: 874–875, 1994.
35) Fadel E, Chapelier AR, Cerrina J. *et al*. Vascular ring causing symptomatic tracheal compression in adulthood. Annales Thorac Surg 5: 1411–1413, 1995.
36) Van Son JAM, Bossert T, Morh FW. Surgical treatment of vascular ring including right cervical aortic arch. J Cardiac Surg 14: 98–102, 1999.
37) Brockes C, Vogt PR, Roth TB, *et al*. Double aortic arch: diagnosis missed for 29 years. Vasa J Vasc Dis 29: 77–79, 2000.
38) Lunde R, Sanders E, Hoskam JAM. Right aortic arch symptomatic in adulthood. Netherlands J Med 60: 212–215, 2002.
39) Greiner A, Perkmann R, Rieger M, *et al*. Vascular ring causing tracheal compression in an adult patient. Ann Thorac Surg 75: 1959–1960, 2003.
40) Chujo M, Miura T, Kawano Y, *et al*. Delayed bronchial stenosis after blunt chest trauma with right aortic arch. Jpn J Thoracic Cardiovascu Surg 52: 78–80, 2004.
41) Kondo C, Takabayashi S, Miyake Y, *et al*. successful surgical treatment for an adult case of double aortic arch. Jpn J Thoracic Cardiovasc Sug 53: 223–226, 2005.
42) Ohhashi Y, Matuo K, Murayama H, *et al*. A case of double aortic arch in adult. J Jpn Soc Vascu Surg. 14(3): PO60 (abstract), 2005.
43) Grillo HC, Wright CD. Tracheal compression with "hairpin" right aortic arch: management by aortic division and aortopexy by right thoracotomy guided by intraoperative bronchoscopy. Ann Thorac Surg 83: 1152–1157, 2007.
44) Bashar AHM, Kazui T, Yamashita K, *et al*. Right aortic arch with aberrant left subclavian artery symptomatic in adulthood. Ann Vascu Surg 20: 529–532, 2006.
45) Joshi AR, Garg A, Vhanmane B, *et al*. A vascular ring variant: an unusual case of vocal cord palsy due to an anomalous left carotid artery arising from a retortracheal arch of the aorta. British J Radiol 79: e81–83, 2006.
46) Lee P, Low S-Y, Liew HL *et al*. Endobronchial ultrasound for detection of tracheomalacia from chronic compression by vascular ring. Respirology 12: 299–301, 2007.
47) Ko Y-K, Hsiao C-L, Lee Y-J. Right aortic arch with a complete vascular ring causing tracheoesophageal compression. J Int Med Taiwan 18: 104–107, 2007.
48) Tjang YS, Aramendi JI, Crespo A, *et al*. Right cervical aortic arch with aberrant left subclavian artery. Asian Cardiovasc Thorac Ann 16: e37–39, 2008.
49) Choh T, Suzuki S, Isomatu Y, *et al*. Total arch replacement for incomplete double aortic arch associated with patent ductus arteriosus in an adult. Ineractive Cardiovasc Thorac Surg 8 269–271, 2009.
50) Sato Y, Tanji M, Yokoyama H. A case of Kommerell's diverticulum with right aortic arch and aberrant left subclavian artery presenting tracheal compression; report of a case. Fukushima Medical J 59(3), 2009.
51) Labelle MF, Rainer WG, Ratzer E, *et al*. Surgical repair of pulmonary artery sling in an adult. Ann Thorac Surg 90: 1009–1011, 2010.