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Anastassios C. Koumbourlis
Mary A. Nevin *Editors*

Pulmonary Complications of Non-Pulmonary Pediatric Disorders

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Editors
Anastassios C. Koumbourlis
George Washington University
School of Medicine
Children's National Medical Center
Washington, DC
USA

Mary A. Nevin
Ann & Robert H. Lurie Children's Hospital
Northwestern Feinberg School of Medicine
Chicago, IL
USA

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Contributors

Alessandro Amaddeo, M.D. Pediatric Noninvasive Ventilation and Sleep Unit, Hôpital Necker Enfants-Malades, Paris, France

Paris Descartes Faculty, Paris, France

Research Unit Inserm U 955, Team 13, Créteil, France

Michael B. Anthracopoulos, M.D. Department of Pediatrics, Pulmonary Division, Medical School of the University of Patras, Rion, Greece

Elinor Charles, L.L.B., M.B.B.S. Division of Asthma, Allergy and Lung Biology, MRC and Asthma UK Centre in Allergic Mechanisms of Asthma, King's College London, London, UK

Caitlin Condit, M.A. Department of Psychology, George Washington University School of Medicine and Health Sciences, Washington, DC, USA

Brigitte Fauroux, M.D., Ph.D. Pediatric Noninvasive Ventilation and Sleep Unit, Hôpital Necker Enfants-Malades, Paris, France

Paris Descartes Faculty, Paris, France

Research Unit Inserm U 955, Team 13, Créteil, France

Adrynne Frazier, M.A. Department of Psychology, George Washington University, School of Medicine and Health Sciences, Washington, DC, USA

Daphna Gelblum, M.D. Department of Radiation Oncology, Memorial Sloan Kettering Cancer Center, West Harrison, NY, USA

Dimos K. Gidaris, M.R.C.P.C.H., M.R.C.P.E., Ph.D. University of Nicosia Medical School, Nicosia, Cyprus

Anne Greenough, M.D.(Cantab), M.B.B.S., D.C.H., F.R.C.P., F.R.C.P.C.H. Department of Women and Children's Health, School of Life Course Sciences, Faculty of Life Sciences and Medicine, King's College London, London, UK

Katie Hunt, M.R.C.P.C.H., M.A.(Cantab), M.B.B.S. Division of Asthma, Allergy and Lung Biology, MRC and Asthma UK Centre in Allergic Mechanisms of Asthma, King's College London, London, UK

Amisha Jain, M.D. Division of Pulmonary and Sleep Medicine, Children's National Health System, George Washington University, School of Medicine and Health Sciences, Washington, DC, USA

Geshani Jayasuriya, B.M.ed., F.R.A.C.P. Department of Respiratory Medicine, The Children's Hospital, Sydney, NSW, Australia

Discipline of Child and Adolescent Health, The University of Sydney, Camperdown, NSW, Australia

Eliot S. Katz, M.D. Division of Respiratory Diseases, Department of Medicine, Children's Hospital, Harvard Medical School, Boston, MA, USA

Steven J. Keogh, M.B.B.S.Hons., F.R.A.C.P. Discipline of Child and Adolescent Health, The University of Sydney, Camperdown, NSW, Australia

Department of Oncology, The Children's Hospital, Sydney, NSW, Australia

Sonia Khirani, Ph.D. Pediatric Noninvasive Ventilation and Sleep Unit, Hôpital Necker Enfants-Malades, Paris, France

ASV Santé, Gennevilliers, France

Anastassios C. Koumbourlis, M.D., M.P.H. Division of Pulmonary and Sleep Medicine, Children's National Medical Center, George Washington University, School of Medicine and Health Sciences, Washington, DC, USA

Geoffrey Kurland, M.D. Division of Pediatric Pulmonology, Children's Hospital of Pittsburgh, Pittsburgh, PA, USA

Donna J. Lee, M.D. Section of Pediatric Pulmonology, Joseph M. Sanzari Children's Hospital, Hackensack University Medical Center, Hackensack, NJ, USA

Nancy Lee, M.D. Department of Radiation Oncology, Memorial Sloan Kettering Cancer Center, New York, NY, USA

Daniel Lewin, Ph.D. Division of Pulmonary and Sleep Medicine, Children's National Health System, George Washington University, School of Medicine and Health Sciences, Washington, DC, USA

Beryl Lin The University of New South Wales, Sydney, NSW, Australia

Sonal Malhotra, M.D. Ann and Robert H. Lurie Children's Hospital of Chicago, Northwestern University Feinberg School of Medicine, Chicago, IL, USA

Chelsea Manchester, M.A. Division of Pulmonary and Sleep Medicine, Children's National Health System, George Washington University, School of Medicine and Health Sciences, Washington, DC, USA

Oscar Henry Mayer, M.D. Division of Pulmonary Medicine, Pulmonary Function Testing Laboratory, The Children's Hospital of Philadelphia, Perelman School of Medicine at the University of Pennsylvania, Philadelphia, PA, USA

Rosemary Megalaa, D.O. Division of Pulmonary and Sleep Medicine, Children's National Health System, School of Medicine and Health Sciences, George Washington University, Washington, DC, USA

Mary A. Nevin, M.D. Ann and Robert H. Lurie Children's Hospital of Chicago, Northwestern University Feinberg School of Medicine, Chicago, IL, USA

R. Paul Boesch, D.O., M.S., F.A.A.P. Mayo Clinic Children's Center, Rochester, MN, USA

Kostas N. Priftis, M.D. Allergy-Pneumonology Department, University of Athens, School of Medicine, Athens, Greece

Thomas G. Saba, M.D. Division of Pediatric Pulmonology, University of Michigan, Ann Arbor, MI, USA

Iman R. Sami-Zakhari, M.D., M.R.C.P. Division of Pulmonary and Sleep Medicine, Children's National Health System, School of Medicine and Health Sciences, George Washington University, Washington, DC, USA

Hiran Selvadurai, M.B.B.S., F.R.A.C.P., Ph.D. Department of Respiratory Medicine, The Children's Hospital, Sydney, NSW, Australia

Discipline of Child and Adolescent Health, The University of Sydney, Camperdown, NSW, Australia

Adam J. Shapiro, M.D. Pediatric Respiratory Medicine, Montréal Children's Hospital, McGill University Health Centre Research Institute, Montréal, QC, Canada

Prateek J. Shukla, M.D. Division of Pulmonary and Sleep Medicine, Children's National Health System, School of Medicine and Health Sciences, George Washington University, Washington, DC, USA

Sylvia Szentpetery, M.D. Division of Pediatric Pulmonology, Children's Hospital of Pittsburgh, Pittsburgh, PA, USA

Daniel J. Weiner, M.D. Division of Pediatric Pulmonology, Children's Hospital of Pittsburgh, Pittsburgh, PA, USA

Pulmonary Complications of Cardiovascular Disorders

Anastassios C. Koumbourlis

Abbreviations

Ao	Aorta
CT	Computed tomography
IVC	Inferior vena cava
LA	Left atrium
LCA	Left carotid artery
LMSB	Left main stem bronchus
LSA	Left subclavian artery
LV	Left ventricle
MEFVC	Maximal expiratory flow-volume curve
MPA	Main pulmonary artery
MRI	Magnetic resonance imaging
RA	Right atrium
RCA	Right carotid artery
RMSB	Right main stem bronchus
RPA	Right pulmonary artery
RSA	Right subclavian artery
RV	Right ventricle
SVC	Superior vena cava

A.C. Koumbourlis, M.D., M.P.H.
Division of Pulmonary and Sleep Medicine, Children's National Medical Center, George Washington University, School of Medicine and Health Sciences, Washington, DC, USA
e-mail: akoumbou@childrensnational.org

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Introduction

The pulmonary complications of the cardiovascular disorders can be categorized into two broad categories: (a) "anatomic," which are usually the result of external compression of the airways by one or more of the large thoracic vessels or by the heart itself (although the bronchioles can also be affected), and (b) "functional" in which the complications are caused by the malfunction of the heart (e.g., congestive heart failure and pulmonary edema).

Normal Anatomic Relationships

The heart is located in the lower mediastinum directly behind the sternum and below the bifurcation of the trachea (Fig. 1). It has a conical shape, with its apex pointing downward and to the left, whereas its base is pointing upward to the right. The heart is "engulfed" by the two lungs which have special grooves to accommodate it. The heart is indirectly connected to both lungs with the right and left branches of the pulmonary artery and the pulmonary veins that enter and exit the lungs in the hilum (Fig. 2a, b). The right atrium (RA) of the heart is located anteriorly to the bronchus intermedius. The left atrium (LA) is adjacent to the left main stem bronchus (LMSB), just before and slightly below the takeoff of the left upper lobe (LUL).

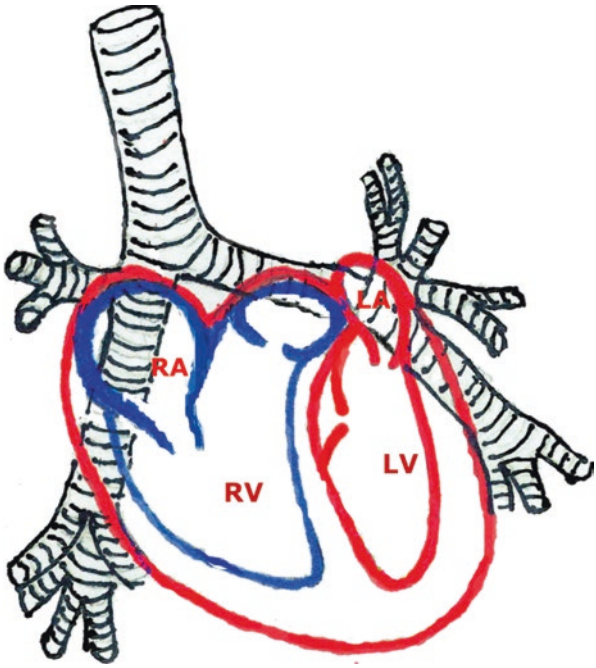

Fig. 1 Schematic representation of the location of the heart in relation to the tracheobronchial tree

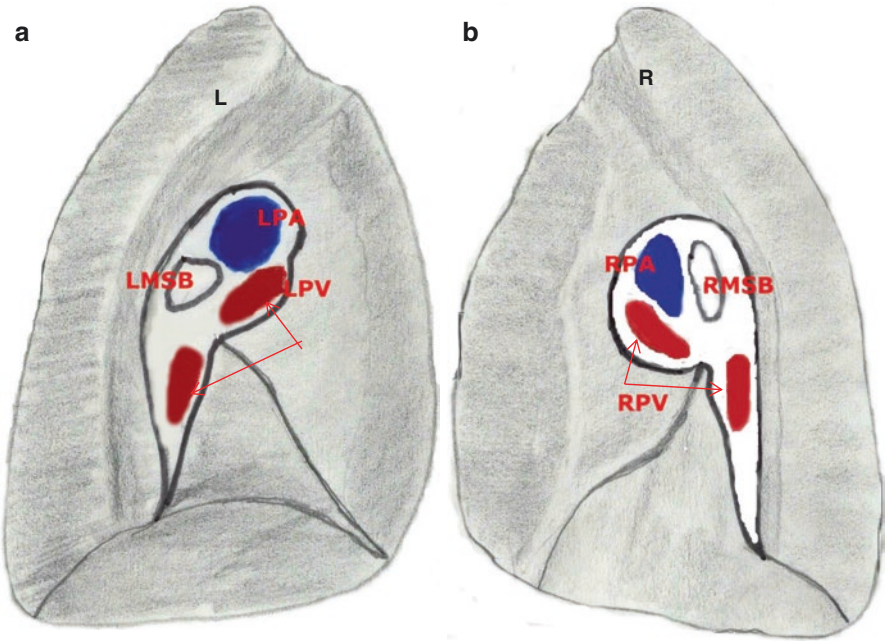

Fig. 2 Schematic representation of the left hilum (**a**) and right hilum (**b**)

Under normal circumstances there is negligible compression of the airways by the atria. In contrast with the heart, many of the big vessels are in very close proximity to the airways causing mild (not clinically significant) compression even under normal circumstances. Specifically:

- *Aorta* (Fig. 3): the *ascending aorta* originates anteriorly and to the right of the trachea. The *aortic arch* follows an oblique course toward the left anterior aspect of the lower trachea near the main carina, slightly compressing it to the right. It then rides over the LMSB and descends posteriorly (*descending aorta*) close to or in contact with the posterior wall of the LMSB. It continues to descend initially to the left and then behind the esophagus and in front of the vertebrae.
- *Brachiocephalic ("innominate") artery* (Fig. 3): it is the first vessel to arise from the aortic arch and transverses upward from left to right. It bifurcates into two branches (*right subclavian and right carotid arteries*). The right common carotid artery courses obliquely very close to the right anterior-lateral aspect of the cervical trachea at the base of the neck (mid-trachea).
- *Main pulmonary artery* (Fig. 4): it originates inferiorly in front of the carina and to the left of the ascending aorta. It branches into the right and left pulmonary arteries.
- *Left pulmonary artery* (Fig. 4): it originates beneath the aortic arch and separates it from the LMSB. It initially courses anterior to the LMSB, but before the takeoff of the LUL, it crosses over, it wraps around the LUL bronchus, and then

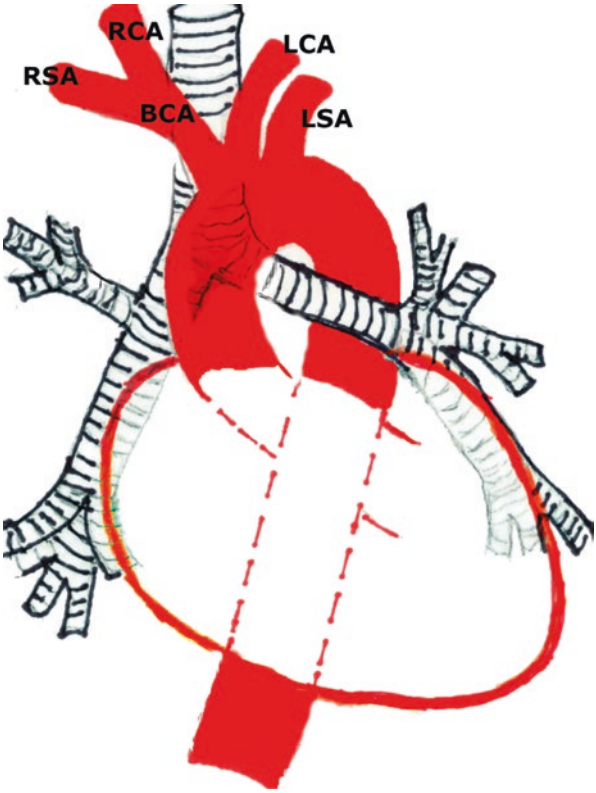

Fig. 3 Schematic representation of the course of the normal aorta and its main branches in relation to the tracheobronchial tree

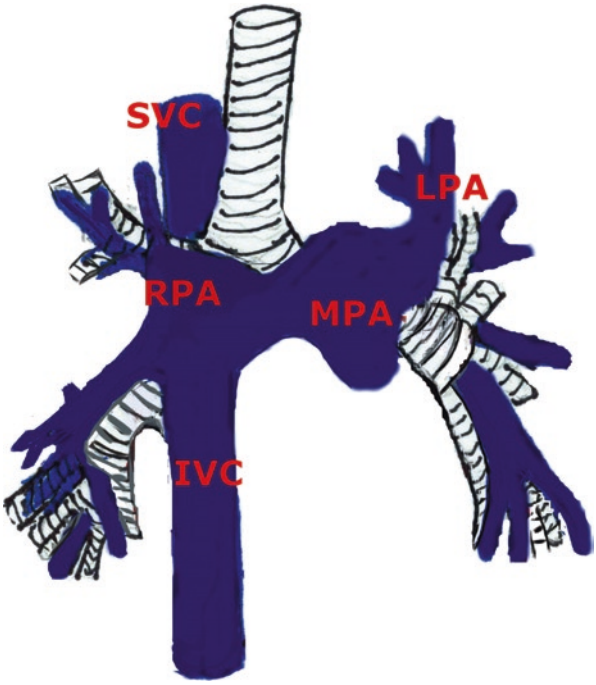

Fig. 4 Schematic representation of the course of the normal main pulmonary artery and its main branches as well as of the superior and inferior vena cava in relation to the tracheobronchial tree

it continues to run anteriorly of the LLL bronchi. In the left hilum, the left pulmonary artery is over the LMSB.
- *Right pulmonary artery* (Fig. 4): it courses almost horizontally in front of the carina, and it is in close contact with the RMSB at its takeoff from the trachea. Its branches remain in close contact with the right upper lobe as well as with the bronchus intermedius and the RML. In the right hilum, the pulmonary artery is directly anterior to the RMSB.
- *Superior vena cava and azygos vein* (Fig. 4): their juncture is in contact with the right anterior aspect of the main carina.
- *Inferior pulmonary veins*: they are located posterior to and are in close contact with the posteromedial aspect of the right and left lower bronchi.
- The *superior pulmonary veins* lie anterior to and inferior to the pulmonary arteries, and they are not in contact with the main stem bronchi.

Under normal circumstances, during bronchoscopy, pulsations and mild compression can be seen in various areas of the tracheobronchial tree, especially in the lower left anterolateral wall of the trachea, on the anterior and posterior wall of the left main stem bronchus, and on the medial segment of the right middle lobe (RML). Significant compression of the lungs causing atelectasis (usually of the left lower lobe) can occur in cases of significant cardiomegaly. Compression of various parts of the tracheobronchial tree occurs when there is enlargement of one or more of the big vessels (e.g., enlargement of the main pulmonary artery and its branches in cases of pulmonary hypertension) and/or in cases of abnormal origin or abnormal course of the vessels.

Abnormal Anatomic Relationships

Cardiomegaly

Most of the typical congenital heart diseases cause initially little or no compression of the lungs or of the airways. Their pulmonary complications tend to be gradual, related either to the development of pulmonary hypertension or of congestive heart failure with pulmonary edema. Notable exceptions are congenital cardiomegaly and Ebstein's anomaly. However, significant cardiomegaly can develop overtime as a result of a host of diverse causes (Table 1). In general, cardiomegaly can develop through one of the four mechanisms: (a) extra volume in one of the heart compartments (usually due to a left-to-right shunt and/or due to valvular malfunction) that allows the regurgitation of blood from the ventricles to the atria, (b) structural "obstruction" of the outflow (e.g., mitral, pulmonary, or aortic valve stenosis), (c) increased afterload that prevents the emptying of the ventricles (e.g., systemic or pulmonary hypertension), and (d) "weakness" of the cardiac muscle itself (e.g., cardiomyopathy) that prevents adequate emptying during systole. These mechanisms may cause enlargement of one or of all the chambers of the heart. Depending on which of the chambers enlarges, the effects will differ. Enlargement of the atria

Table 1 Conditions causing cardiomegaly

Enlargement of the entire heart	
Primary mechanism: conditions affecting the cardiac muscle	• Congenital cardiomegaly • Idiopathic cardiomyopathy • Dilated cardiomyopathy • Hypertrophic cardiomyopathy
Enlargement of the right side of the heart	
Primary mechanisms: 1. Increase in the blood volume in the right atrium and/or ventricle	• Atrial septal defect (ASD) • Ventricular septal defect (VSD) • Tricuspid regurgitation • Anomalous pulmonary venous return • Tetralogy of Fallot *Systemic noncardiac conditions* – Athletic heart – Pregnancy – Renal failure – Anemia – Scleroderma – Systemic lupus erythematosus – Rheumatoid arthritis
2. Right-sided outflow obstruction	• Tricuspid stenosis • Pulmonary valve stenosis • Cor pulmonale • Pulmonary arterial hypertension
Enlargement of the left side of the heart	
Primary mechanisms: 1. Increase in the blood volume in the left atrium and/or ventricle	• Mitral regurgitation • Aortic regurgitation
2. Left-sided outflow obstruction	• Mitral stenosis • Aortic stenosis • Hypertension

is much more likely to cause compression of the main stem bronchi. Enlargement of the left atrium may actually push the left main stem upward in a more horizontal position. In contrast, enlargement of the ventricles tends to compress the lungs causing atelectasis.

Congenital Vascular Abnormalities: "Rings, Slings, and Other Things"

Various congenital vascular abnormalities do cause direct compression of the trachea and/or of the bronchi. The vast majority of these abnormalities consist of an abnormal aortic arch, in combination with a left (or less commonly a right) ligamentum arteriosum and/or an aberrant subclavian artery, which create a "ring" formation around the trachea. The most common of the aortic arch abnormalities are the double aortic arch and the right aortic arch. The ligamentum arteriosum is the remnant of the ductus arteriosus that normally disappears during the first 2–3 weeks of

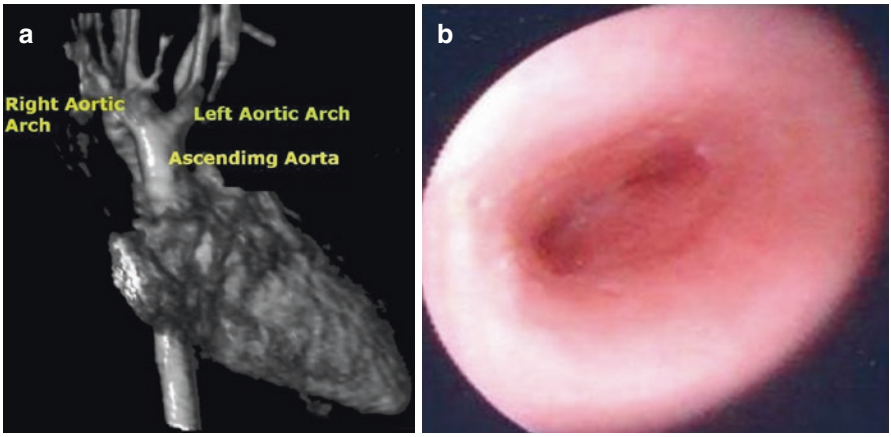

Fig. 5 3-D reconstruction from a cardiac MRI showing the double aortic arch (**a**); severe compression of the lower trachea due to a complete vascular ring (**b**)

life. When it fails to involute, it becomes a small ligament that connects the right aortic arch (or of one of its branches) to the left pulmonary artery.

Double Aortic Arch (Fig. 5)

- *Frequency*: one of the two most common vascular abnormalities.
- *Components*: it is caused by the failure of the fourth right arch to involute. As a result there are two arches (left and right) that are both connected to the descending aorta.
- *Special features*: it encircles the trachea and the esophagus; in 30% of the cases, the smaller arch is atretic; it is usually not associated with intracardiac defects.
- *Clinical features*: it usually causes symptoms (harsh inspiratory and expiratory "wheezy" sound) early in life (even at birth) that tend to become worse after the first few weeks as the infant becomes more acting. However, if it is not very tight, it can remain undiagnosed for years and may manifest itself later in life as "exercise intolerance."

Right Aortic Arch

- *Frequency*: rare in the general population but common among patients with other intracardiac anomalies (especially patients with tetralogy of Fallot).
- *Components*: a "ring" is being formed by the aortic arch that is located on the right side of the trachea, the ascending aorta anteriorly, the descending aorta posteriorly, and the ligamentum arteriosum and left pulmonary artery on the left.
- *Special features*: the ring encircled the trachea and the esophagus; it is associated with intracardiac defects in 10% of the cases.
- *Clinical features*: it causes symptoms early in life.

Right Aortic Arch with Aberrant Left Subclavian Artery and Left Ligamentum Arteriosum

- *Frequency*: rare in the general population.
- *Components*: in this formation the right arch gives off both the left and the right carotid arteries and the right and left subclavian arteries. The "ring" is formed by the right arch to the right, the left carotid artery that travels across the anterior wall of the trachea, and the left subclavian artery that courses from right to left, and it is completed by the left ligamentum arteriosum that connects the left subclavian with the left pulmonary artery.
- *Special features*: the ring encircled the trachea and the esophagus; it is associated with intracardiac defects in 10% of the cases.
- *Clinical features*: it causes symptoms early in life.

Right Aortic Arch with Mirror-Image Branching and Retroesophageal Ligamentum Arteriosum

- *Frequency*: rare in the general population.
- *Formation of the ring*: the left innominate artery is the first branch of the right arch, and it then branches into the left carotid and left subclavian arteries, both of whom are branches of the left innominate artery and course over the anterior tracheal wall. The right carotid artery is the second branch of the aortic arch followed by the right subclavian artery and finally the ligamentum arteriosum that originates from the Kommerell diverticulum that is the remnant of the left fourth arch, and it is located at the point where the right arch merges with the descending thoracic aorta. The ligamentum crosses to the left behind the esophagus and then travels anteriorly where it completes the ring when it joins the left pulmonary artery. Often the ligamentum originates from the left innominate artery of left subclavian artery, so it does not form a complete ring.
- *Special features*: this type of anomaly is associated with intracardiac defects in up to 90% of the cases.

Left Aortic Arch with Right Descending Aorta and Right Ligamentum Arteriosum

- *Frequency*: extremely rare.
- *Formation of the "ring"*: in this variant the first branch of the left aortic arch is the right common carotid artery that crosses to the right over the anterior tracheal wall. The next vessel is the left carotid, followed by the left subclavian artery. The right subclavian artery is branching off the proximal right-sided descending aorta. The ligamentum arteriosum branches off the base of the right subclavian (or from a nearby diverticulum) and connects it to the right pulmonary artery.

Left Aortic Arch, Right Descending Aorta, and Atretic Right Aortic Arch

- *Frequency*: very rare.
- *Formation of the "ring"*: the brachiocephalic vessels are branching normally of the left aortic arch. However, the arch travels behind the esophagus and joins the descending aorta that is right sided. The "ring" is completed by an atretic right arch.

Tracheal Compression by Vascular Structures but Without Formation of a Complete Ring

Anomalous Brachiocephalic (Innominate) Artery (Fig. 6)

- *Frequency*: it is the most common type of tracheal compression.
- *Anatomy*: the innominate artery originates from the brachiocephalic artery that originates in the left aortic arch and crosses normally over the anterior wall of the trachea from left to right. In certain cases it is originating more distally, and thus it crosses much closer to the tracheal wall than usual.
- *Special features*: tracheal compression by the innominate artery is very easily recognizable during bronchoscopy because it causes a characteristic compression of the right anterior-lateral tracheal wall. It is more difficult to diagnose radiographically or angiographically because the course of the artery is normal.

Retroesophageal Right Subclavian Artery with Left Aortic arch and Left Ligamentum Arteriosum

- *Frequency*: very common, occurring in approximately 0.5% of the general population.
- *Anatomical features*: in this variant the right subclavian artery originates from the descending aorta and courses posterior to the esophagus. The left ligamentum arteriosus originates from the aortic arch and connects to the left pulmonary artery.

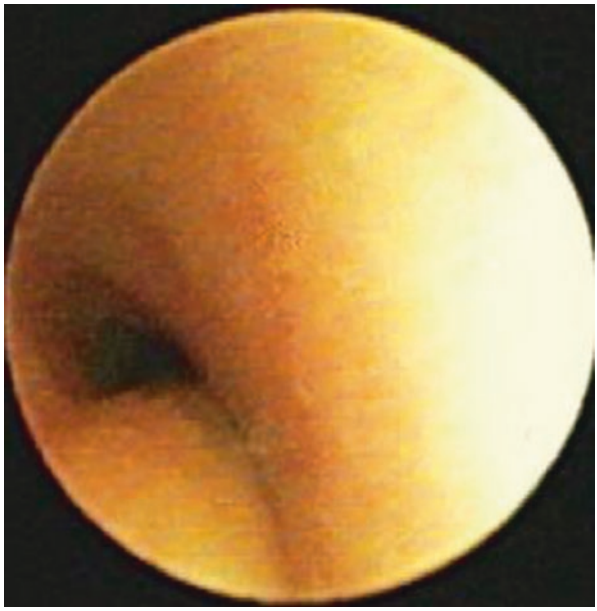

Fig. 6 Bronchoscopic picture showing external compression of the anterior right wall of the mid-trachea by the innominate artery

- *Special features*: this variant does not usually cause respiratory or other symptoms. However, depending on the degree of compression of the esophagus, it may cause symptoms of dysphagia.

Left Pulmonary Artery Sling (Fig. 7)

- *Frequency*: It is estimated to account for approximately 10% of the non-aortic arch-related vascular compressions.
- *Anatomical features*: in this pathologic variant, the left pulmonary artery originates from the right pulmonary artery and crosses over the right main stem bronchus and circles the trachea at its bifurcation and then crosses to the left in between the trachea and the esophagus.
- *Special features*: it causes a characteristic indentation of the right lower wall just above the bifurcation that is visible both bronchoscopically and radiographically. Pulmonary artery slings are often associated with tetralogy of Fallot as well as with tracheal stenosis with complete tracheal rings. In such cases the trachea and the main stem bronchi have a characteristic appearance of an inverted capital "T" due to the virtually horizontal position of the main stem bronchi.

*Scimitar Syndrome (Pulmonary Venolobar Syndrome) (*Fig. 8*)*

- *Frequency*: rare.
- *Anatomical features*: partial anomalous pulmonary venous return from the right lung to the inferior vena cava (usually near its junction with the right atrium).
- *Special features*: the anomalous pulmonary vein has a curved appearance that resembles the curved sword known as "scimitar." The syndrome is associated with right lung hypoplasia (including a hypoplastic right pulmonary artery) that displaces the heart to the right hemithorax.

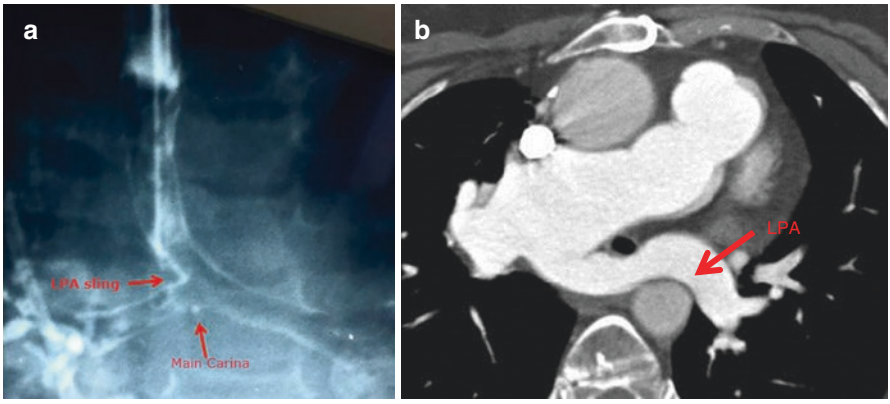

Fig. 7 **(a)** Bronchographic image showing the indentation of the right tracheal wall just above the carina caused by a left pulmonary artery sling. Note the almost horizontal position of the two main stem bronchi that give the appearance of an inverted capital T; (**b**) CT scan showing left pulmonary artery sling compressing the trachea and the esophagus

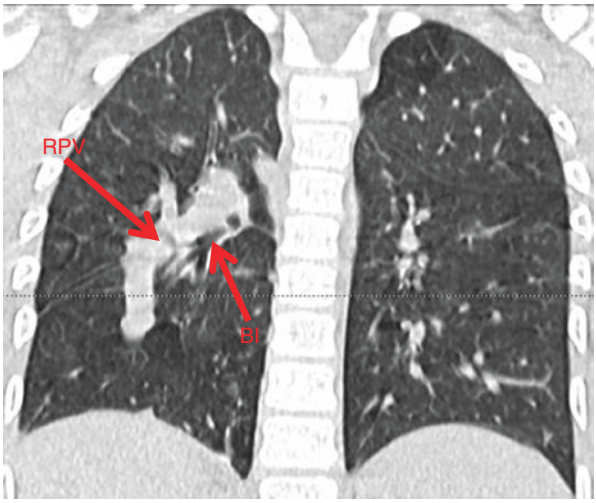

Fig. 8 Chest CT showing a curved density resembling a "scimitar" sword, formed by the abnormal course of the right pulmonary vein

Clinical Presentation of Tracheobronchial Compression by Vascular Structures

Onset of symptoms: it is variable. Severe compression will produce symptoms shortly after birth, whereas mild compression may remain asymptomatic (and undiagnosed) for years. Tracheal stenosis with complete tracheal rings (usually associated with pulmonary artery sling) tends to get progressively worse as the infant grows because the tracheal diameter does not increase despite the growth of the rest of the body. A common feature among virtually all vascular abnormalities is that they impair the clearance of secretions from the compressed airways (especially when it is associated with tracheomalacia). The retention of secretions promotes the colonization of the airways usually with bacterial organisms, and the patients often have manifestations of chronic bronchitis and/or recurrent pneumonias.

Clinical Features

Chest wall retractions: suprasternal chest wall retractions tend to be a standard feature of extrathoracic and large intrathoracic obstruction even in the absence of respiratory distress. However, they can be easily missed in young infants because their chin tends to cover the suprasternal notch. When the obstruction is significant, the patients have substernal and intercostal retractions.

Cough: because compression of the trachea usually produces a certain degree of tracheomalacia, affected patients develop a very characteristic dry cough with a "honking" quality ("goose honking"). When there are secretions, the cough is congested, hacking often described by the parents as "smokers' cough."