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Compression of Adjacent Anatomical Structures by Pulmonary Artery Dilation

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Abstract

Pulmonary hypertension is the commonest condition leading to dilated pulmonary artery. We describe three different types of compression of adjacent anatomical structures by dilated pulmonary arteries. We included involvement of the left main coronary artery, left recurrent laryngeal nerve and tracheobronchial tree. Compression of these structures can cause major complications such as myocardial ischemia, hoarseness and major airway stenosis. We present a case for each scenario and review the literature for each of these complications, focusing on patients' characteristics and contemporary management.

Keywords

Dilated pulmonary artery; left main coronary artery; left recurrent laryngeal nerve; tracheobronchial tree; pulmonary hypertension

Introduction

Pulmonary arterial hypertension (PAH) is a progressive disease defined by abnormally elevated pulmonary pressures that can lead to right ventricular (RV) failure and death [1]. The persistent elevation of the pulmonary artery pressure leads to vascular remodeling resulting in dilation of pulmonary arteries [2]. Pulmonary artery dilation is increasingly recognized in PAH patients and this enlarged main vessel could impinge upon adjacent anatomical structures such as the left main coronary artery (LMCA), left recurrent laryngeal nerve (LRLN), and less commonly, tracheobronchial tree [3-5]. Compression of these structures can cause major complications such myocardial ischemia and even sudden death, hoarseness and major airway stenosis. These life threatening manifestations commonly remain under recognized and symptoms may be attributed to the natural course of the

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disease [2]. Advances in imaging techniques, i.e. computed tomography (CT) or magnetic resonance imaging (MRI) have allowed the identification of pulmonary artery dilation and its potential influence over adjacent anatomical structures. We describe three different types of compression of adjacent structures by dilated pulmonary arteries, including compression of the LMCA, LRLN and tracheobronchial tree. We present a case for each scenario and review the literature for each of these complications, focusing on patient characteristics and contemporary management.

Case presentations

a) Left main coronary artery compression

A 65-year-old woman with chronic thromboembolic pulmonary hypertension underwent pulmonary thromboendarterectomy two decades before her current presentation. Due to residual pulmonary hypertension (PH), she had been treated with intravenous treprostinil. She was referred to our center for consideration of heart-lung transplant. Her medical history included a positive lupus anticoagulant and recurrent pulmonary embolism. During her hospital stay she experienced an acute episode of chest pain with anterolateral ST segment depression on the electrocardiogram and elevated cardiac enzymes, consistent with non-ST elevation myocardial infarction. Chest XR showed markedly dilated pulmonary arteries (Figure 1, panel A). Echocardiography revealed normal left and right ventricular (RV) systolic function with an estimated RV systolic pressure of 71 mm Hg and markedly dilated main pulmonary artery measuring 8 cm in diameter. Right heart catheterization (RHC) showed a right atrial (RA) pressure of 13 mm Hg, mean pulmonary artery pressure (mPAP) of 55 mm Hg, pulmonary artery wedge pressure (PAWP) of 19 mm Hg, cardiac output of 4.7 L/min and pulmonary vascular resistance (PVR) of 7.7 Wood units.

Coronary angiography revealed 80% proximal narrowing of the LMCA (Figure 1, panel B and D) which was confirmed by intravascular ultrasound (intravascular area of 5 mm²). A cardiac computed tomography showed enlarged pulmonary arteries (main pulmonary artery diameter: 9.5 cm, main pulmonary artery to aortic diameter ratio (PA/Ao ratio): 2.6, right pulmonary artery diameter: 6.9 cm and left pulmonary artery diameter: 3.8 cm) with extrinsic compression by the right pulmonary artery of the proximal LMCA and proximal bronchi at the level of the carina (Figure 1, panel C and E). Beside optimization of PH-specific treatment, the patient underwent percutaneous coronary intervention with the delivery of an 18 mm drug-eluting stent. She tolerated the procedure well and had no residual stenosis; however, the patient died 9 months after the intervention from RV failure.

b) Left recurrent nerve compression

A 42-year-old nonsmoker woman with idiopathic PAH and right heart failure presented to our institution for lung transplant evaluation. She had been treated with epoprostenol, sildenafil and warfarin. She experienced worsening of her dyspnea and hoarseness of her voice. On examination she had a high pitched voice. Fiberoptic laryngoscopy revealed left vocal cord paralysis without evidence of any anatomical lesion. Echocardiography showed severely decreased RV systolic function with an estimated RV systolic pressure of 97 mm Hg. RHC revealed a RA pressure of 8 mm Hg, mPAP of 45 mm Hg, PAWP of 8 mm Hg,

cardiac output of 2.4 L/min and PVR of 15.4 Wood units. A contrast enhanced computed tomography of the chest and neck depicted enlarged pulmonary arteries (main pulmonary artery pulmonary diameter of 3.5 cm with a PA/Ao ratio of 1.28). There were no lung nodules or enlarged lymph nodes in the chest. Nevertheless, the enlarged pulmonary artery impinged the LRLN causing left vocal cord paralysis (Figure 2). The PAH-specific treatment was optimized but the patient suddenly died two years after this complication.

c) Tracheobronchial tree compression

A 48-year-old woman with history of complex cyanotic congenital heart disease (CHD) that comprised double outlet RV, pulmonic valve stenosis, subaortic ventricular septal defect and dextrocardia. During her childhood she underwent Blalock-Taussig and Waterston shunts. She had been treated with bosentan and tadalafil for her Eisenmenger syndrome. The enlarged pulmonary artery caused compression of her main bronchus with development of right lower lobe bronchiectasis. She presented to our institution with progressive dyspnea due to an exacerbation of her bronchiectasis.

Given her declining clinical condition she was evaluated for heart and lung transplant. Chest XR showed dextrocardia with dilated pulmonary arteries (Figure 3, panel A).

Echocardiography revealed a large ventricular septal defect and severely dilated double-outlet RV with severe systolic dysfunction. RHC showed a RA pressure of 7 mm Hg, mPAP of 93 mm Hg, PAWP of 9 mm Hg, cardiac output of 4.1 L/min, PVR of 20.5 Wood units with Qp/Qs is 0.5:1. Computed tomography of the chest showed massive dilation of the central pulmonary arteries causing significant narrowing of the distal trachea and proximal main stem bronchi (Figure 3, panel B) as well as distal bronchiectasis of the right lower lobe bronchi (Figure 3, panel C). Magnetic resonance angiography showed a main pulmonary artery of 3.9 cm, PA/Ao ratio of 1.3, right and left pulmonary arteries of 5.4 cm and 4.8 cm, respectively (Figure 3, panel D, E and F).

Intravenous treprostinil was started in an attempt to reduce her pulmonary artery pressure and wall tension. Interventional pulmonology considered that a stent placement would not be appropriate, given concern for localized tissue hypoperfusion and erosion of the stent into the pulmonary artery. Her intensive care unit course was complicated by septic shock that caused her death. Autopsy revealed severely dilated and calcified pulmonary arteries, plexiform lesions, and extrinsic compression of the right main bronchus by the dilated right pulmonary artery.

Discussion

We presented three cases in which an enlarged pulmonary artery due to PH was responsible for compression of a) LMCA leading to an acute coronary syndrome, b) LRLN causing hoarseness and c) tracheobronchial tree with development of localized bronchiectasis. These complications portend a poor prognosis since they reflect an advanced stage of the disease. In fact, the three patients presented in this manuscript died shortly after noticing the compressions of adjacent anatomical structures. Hence we consider of relevance to raise awareness on these life-threatening conditions to prompt an earlier diagnosis and aggressive PH treatment.

a) Left main coronary artery compression

A dilated pulmonary artery can compress the LMCA and when this vessel reaches a critical narrowing, left ventricular ischemia ensues. This complication clinically presents with exertional chest pain [6] and less commonly syncope [7], myocardial infarction or sudden death [8 9]. Of note is that chest pain is reported by over 40% of PAH patients [10] and is typically caused by RV hypoperfusion and demand ischemia resulting from increased RV load [11], pulmonary artery distention with activation of pain receptors [12] and less commonly by LMCA compression by a dilated pulmonary artery. This latter complication is more frequently seen in patients with long standing PAH [5 13] and particularly in subjects with idiopathic PAH [5 7 14], PAH associated with atrial septal defect [13] and chronic thromboembolic pulmonary hypertension [15 16] (Table 1).

The risk of LMCA compression depends on the site of origin of this vessel in the aorta. For instance, a left coronary sinus that is inferiorly displaced and/or rightward positioned is more prone to compression from a dilated pulmonary artery. Conversely, other locations of the left coronary sinus might be protective and this explains why some patients with very large pulmonary arteries do not develop this complication [17 18]. Furthermore, younger age and female gender appear to be risk factors [13]. However, Mesquita et al [13] found no association between age, gender, PAH etiology and LMCA compression. These authors noted that patients with LMCA impingement had significantly larger main pulmonary artery diameters (mean $\pm$ standard deviation of 5.5 ± 1.3 cm) and PA:Ao diameter ratios (1.98 ± 0.55). Remarkably, they observed LMCA compression only when the main pulmonary artery diameter was > 4.0 cm and PA:Ao ratio > 1.21 .

Computed tomography angiography and magnetic resonance imaging are the preferred initial screening tests for LMCA compression [9]. Coronary angiography preferentially with intravascular ultrasound (IVUS) remains the gold standard for the diagnosis since it can distinguish extrinsic compression, characterized by smooth narrowing of the LMCA, from intrinsic atherosclerosis [5 19]. In addition, the degree of LMCA narrowing can be further evaluated by IVUS and fractional flow reserve to help determine the need for revascularization [20]. Remarkably, a limited amount of atherosclerotic plaque in the LMCA can significantly reduce the intravascular area in the context of marked extravascular compression [19].

Importantly, PAH-specific therapies can worsen the myocardial ischemia and left ventricular function [9 14 21 22] probably because of subendocardial-subepicardial steal [23]. Definitive treatment of LMCA compression by an enlarged pulmonary artery involves percutaneous coronary intervention (PCI) with stent placement [14], coronary artery bypass grafting (CABG) or lung or lung-heart transplantation [9]. Importantly, these interventions need to be performed in highly specialized centers given the high risk of complications.

We identified a total of 54 patients with PAH and compression of the LMCA in the literature (Table 1). Age ranged from 16 to 67 years. Most of the patients were women ($n = 37$, 69 %), etiology of PH was predominantly PAH-associated with CHD ($n = 37$, 69 %) and idiopathic PAH ($n = 12$, 22 %). PAH-specific therapy was provided to 16 patients (30 %). Time from PAH diagnosis and LMCA compression was only reported in 16 patients and ranged from

0.5 to 27 years. The degree of extrinsic LMCA stenosis fluctuated from 50 to 95 %. The size of the pulmonary artery and mean PAP varied from 3.5 to 12.0 cm and 30 to 127 mmHg, respectively. Treatment provided included PCI with stent placement (n= 22, 41 %) or surgical revascularization (n= 9, 17 %).

b) Left recurrent nerve compression

Anatomically, the left vagus nerve gives rise to the LRLN at the level of the aortic arch. The LRLN loops posteriorly under the aortic arch and travels towards the neck in the groove between the esophagus and trachea, to finally innervate the intrinsic muscles of the larynx that control the left vocal cord. The relative long course of the LRLN makes it susceptible to compression [24]. A cardiovocal syndrome is defined by LRLN disruption with vocal cord palsy and hoarseness secondary to a cardiovascular cause. This syndrome was first described by Ortner in 1897 in two patients with mitral valve stenosis and vocal cord paralysis. Ortner initially attributed the LRLN compression to a dilated left atrium [3]. Since that time many theories emerged to explain the cardiovocal syndrome [25] including a LRLN 'fixation' by lymphadenopathy or scarring in the aortic window, bronchial compression, atherosclerosis of the pulmonary artery and impingement by the ligamentum arteriosum or dilated pulmonary artery.

Autopsy studies showed that the distance between the pulmonary artery and the aorta, at the level of the aortic window, is as small as 0.4 cm, and therefore the LRLN is likely compressed between the pulmonary artery and aorta [3]. Left vocal cord palsy due to LRLN compression was reported in patients with idiopathic PAH [3 26-28], PAH due to CHD [26 29-31] and chronic thromboembolic pulmonary hypertension [24 32]. Interestingly, in several cases, the hoarseness improved upon reducing the pulmonary artery pressure with PAH-specific therapies [3 25 33] (Table 2). Indirect laryngoscopy is essential to confirm diagnosis and to rule out other causes of left vocal cord palsy. The origin of the LRLN compression should be investigated by obtaining computed tomography and/or magnetic resonance imaging of the neck and chest [24 25].

Upon reviewing the literature we found a total of 25 patients (Table 2) that experienced compression of the LRLN due to an enlarged pulmonary artery. Of the 15 patients in whom age and gender were provided, 8 (53%) were men and their age ranged from 19 to 56 years. Idiopathic PAH (n= 19, 76 %) and PAH-associated with CHD (n= 4, 16%) were the two most frequent etiologies of PH. The time from PAH diagnosis and LMCA compression was reported in 8 patients and it extended from 1 to 60 months. Hoarseness was the presenting symptoms in 16 (64 %) out of the 25 patients. The mean PAP varied from 29 to 101 mmHg. Remarkably, three patients (50 %) out of 6 who received PAH-specific therapies improved their hoarseness.

c) Tracheobronchial tree compression

Given the vicinity of the pulmonary artery to the tracheobronchial tree, compression of the airways by an enlarged pulmonary artery is certainly possible. Tracheobronchial tree compression is more frequently reported in children, because younger individuals have more elastic airways which are more susceptible to compression [4]. Therefore the predominant

type of PH that leads to this complication is PAH associated with CHD [34]. In PAH associated with CHD, the pulmonary artery dilates because of the increase flow and shear stress, hypoxemia, vascular atherosclerosis and/or inherited defects that debilitate the medial vascular layer [35]. Medical and surgical advances have allowed patients with complex CHD patients reach adulthood [35], allowing more time for the pulmonary artery to dilate. The usual presentation of a tracheobronchial tree compression includes persistent wheezing and cough, symptoms that can mislead the diagnosis to asthma or respiratory infections [30 36] (Table 3).

There are four sites in the tracheobronchial tree which are at a higher risk for compression by a dilated pulmonary artery [37]. The most common site is where the left pulmonary artery crosses the superior aspect of the left main bronchus. The second site is where the left pulmonary artery arches over the origin of left upper bronchus. The third site is at the point where the right pulmonary artery crosses the anterior aspect of the bronchus intermedius when it gives rise to the right middle bronchus. The last compression site is at the left tracheal border, where the aorta is pushed superiorly and medially by an enlarged left pulmonary artery.

Computed tomography angiography and magnetic resonance imaging of the chest are useful modalities to assess potential airway stenosis caused by the pulmonary vasculature [34 36]. Flexible bronchoscopy is usually not needed for this diagnosis and the procedure may pose an increased risk in patients with severe PH [38]. Pulmonary arterial hypertension-specific therapies can reduce the intravascular pressure but as the vascular radius would not decrease, the wall tension would remain elevated. Airway stents can also generate tension that may decrease perfusion of the bronchial wall, and with time, erode into the surrounding tissues towards the adjacent vasculature [39 40]. Surgical interventions to relieve the extrinsic airway compression are of high risk in patients with PH [41]. Therefore, at this point there are no effective treatments for this condition and it is unclear whether early initiation of PAH-specific therapies particularly in patients with CHD-associated PAH, may prevent the enlargement of the pulmonary arteries and related complications.

We were able to identify in the literature, six adult patients with PAH and compression of the bronchial airway (Table 3). In the five patients in whom data were given, the age extended from 24 to 56 years and the majority were men (n= 4, 80 %). As expected, PAH-associated with CHD (n= 5, 83 %) was the most frequent type of PH associated with this complication. The left and right main stem bronchi were the predominant sites of airway compression, and PAH-specific therapy was provided to half of the patients (3 patients) with unclear benefits.

d) Combined extrinsic compression

Compression of more than one adjacent anatomical structure, like in our first case, has been previously reported. Achouh et al. [30] presented two cases of airway stenosis due to extrinsic compression from an enlarged pulmonary artery. One of these patients also had LMCA compression and the other left vocal cord palsy. Andjelkovic et al. [29] described a patient with compression of both LMCA and LRLN. Similarly, Kawut et al [5] presented 3 patients with LMCA compression by enlarged pulmonary arteries and one of these patients also suffered from hoarseness.

e) Prognostic implications of a dilated pulmonary artery

A dilated pulmonary artery can compress adjacent anatomical structures and also lead to pulmonary artery dissection and rupture [42]. Pulmonary artery dissections usually occur at the level of the main pulmonary artery that can then rupture into the pericardium or pleural cavity [43]. Commonly, pulmonary artery dissection is a post-mortem diagnosis [44]; however a few cases were diagnosed alive and underwent successful repair of the pulmonary artery [42].

A dilated pulmonary artery was shown to be an independent risk factor of sudden death in patients with PAH [45], as a result of pulmonary artery dissection / rupture or compression of the LMCA leading to ischemia and lethal arrhythmias [45]. Therefore, in the appropriate setting it may be valuable to determine the pulmonary artery size and assess the risk of compression of adjacent anatomical structures, particularly the LMCA. We have previously described an algorithm to assist clinicians in evaluating a dilated pulmonary artery in adult patients [2]. Further investigations are needed to determine the value of regularly monitoring the pulmonary artery size in subjects with long standing PAH.

Conclusions

An enlarged pulmonary artery can lead to extrinsic compression of the surrounding vital structures such as the LMCA, LRLN and tracheobronchial tree. The diagnosis requires a high index of suspicion since these complications can be mistaken by more common pathologies. These local complications usually entail a poor prognosis since they reflect an advanced stage of the disease. It remains unclear whether treatment with PAH-specific therapies would improve or prevent these complications. Due to the poor prognosis and limited therapeutic options, these patients need management by multidisciplinary teams in highly specialized centers.

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References

1. Tonelli AR, Arelli V, Minai OA, et al. Causes and circumstances of death in pulmonary arterial hypertension. *Am J Respir Crit Care Med*. 2013; 188(3):365–9. doi: 10.1164/rccm.201209-1640OC[published Online First: Epub Date]]. [PubMed: 23600433]
2. Raymond TE, Khabbaza JE, Yadav R, et al. Significance of main pulmonary artery dilation on imaging studies. *Ann Am Thorac Soc*. 2014; 11(10):1623–32. doi: 10.1513/AnnalsATS.201406-253PP[published Online First: Epub Date]]. [PubMed: 25406836]
3. Shankar O, Lohiya BV. Cardiovascular syndrome--a rare presentation of primary pulmonary hypertension. *Indian heart journal*. 2014; 66(3):375–7. doi: 10.1016/j.ihj.2013.12.055[published Online First: Epub Date]]. [PubMed: 24973850]
4. Edwards JE, Burchell HB. Effects of pulmonary hypertension of the tracheobronchial tree. *Diseases of the chest*. 1960; 38:272–84. [PubMed: 13819416]

5. Kawut SM, Silvestry FE, Ferrari VA, et al. Extrinsic compression of the left main coronary artery by the pulmonary artery in patients with long-standing pulmonary hypertension. *The American journal of cardiology*. 1999; 83(6):984–6, A10. [PubMed: 10190427]
6. Lee MS, Oyama J, Bhatia R, et al. Left main coronary artery compression from pulmonary artery enlargement due to pulmonary hypertension: a contemporary review and argument for percutaneous revascularization. *Catheterization and cardiovascular interventions : official journal of the Society for Cardiac Angiography & Interventions*. 2010; 76(4):543–50. doi: 10.1002/ccd.22592[published Online First: Epub Date]. [PubMed: 20506194]
7. Gomez Varela S, Montes Orbe PM, Alcibar Villa J, et al. [Stenting in primary pulmonary hypertension with compression of the left main coronary artery]. *Revista espanola de cardiologia*. 2004; 57(7):695–8. [PubMed: 15274855]
8. Vaseghi M, Lee JS, Currier JW. Acute myocardial infarction secondary to left main coronary artery compression by pulmonary artery aneurysm in pulmonary arterial hypertension. *The Journal of invasive cardiology*. 2007; 19(12):E375–7. [PubMed: 18180532]
9. Lindsey JB, Brilakis ES, Banerjee S. Acute coronary syndrome due to extrinsic compression of the left main coronary artery in a patient with severe pulmonary hypertension: successful treatment with percutaneous coronary intervention. *Cardiovascular revascularization medicine : including molecular interventions*. 2008; 9(1):47–51. doi: 10.1016/j.carrev.2007.07.003[published Online First: Epub Date]. [PubMed: 18206638]
10. Rich S, Dantzker DR, Ayres SM, et al. Primary pulmonary hypertension. A national prospective study. *Annals of internal medicine*. 1987; 107(2):216–23. [PubMed: 3605900]
11. Vlahakes GJ, Turley K, Hoffman JI. The pathophysiology of failure in acute right ventricular hypertension: hemodynamic and biochemical correlations. *Circulation*. 1981; 63(1):87–95. [PubMed: 7438411]
12. Viar WN, Harrison TR. Chest pain in association with pulmonary hypertension; its similarity to the pain of coronary disease. *Circulation*. 1952; 5(1):1–11. [PubMed: 14896447]
13. Mesquita SM, Castro CR, Ikari NM, et al. Likelihood of left main coronary artery compression based on pulmonary trunk diameter in patients with pulmonary hypertension. *The American journal of medicine*. 2004; 116(6):369–74. doi: 10.1016/j.amjmed.2003.11.015[published Online First: Epub Date]. [PubMed: 15006585]
14. Rich S, McLaughlin VV, O'Neill W. Stenting to reverse left ventricular ischemia due to left main coronary artery compression in primary pulmonary hypertension. *Chest*. 2001; 120(4):1412–5. [PubMed: 11591592]
15. Bonderman D, Fleischmann D, Prokop M, et al. Images in cardiovascular medicine. Left main coronary artery compression by the pulmonary trunk in pulmonary hypertension. *Circulation*. 2002; 105(2):265. [PubMed: 11790711]
16. Ngaage DL, Lapeyre AC, McGregor CG. Left main coronary artery compression in chronic thromboembolic pulmonary hypertension. *European journal of cardio-thoracic surgery : official journal of the European Association for Cardio-thoracic Surgery*. 2005; 27(3):512. doi: 10.1016/j.ejcts.2004.10.060[published Online First: Epub Date]. [PubMed: 15740965]
17. Fujiwara K, Naito Y, Higashiue S, et al. Left main coronary trunk compression by dilated main pulmonary artery in atrial septal defect. Report of three cases. *The Journal of thoracic and cardiovascular surgery*. 1992; 104(2):449–52. [PubMed: 1495309]
18. Kajita LJ, Martinez EE, Ambrose JA, et al. Extrinsic compression of the left main coronary artery by a dilated pulmonary artery: clinical, angiographic, and hemodynamic determinants. *Catheterization and cardiovascular interventions : official journal of the Society for Cardiac Angiography & Interventions*. 2001; 52(1):49–54. [PubMed: 11146522]
19. Demkow M, Kalinczuk L, Kepka C, et al. Left main artery compression by pulmonary artery aneurysm and ostial athero-stenosis of left anterior descending artery in a young female with pulmonary arterial hypertension. *Eur Heart J*. 2012; 33(20):2621. doi: 10.1093/eurheartj/ehs195[published Online First: Epub Date]. [PubMed: 22782942]
20. Pina Y, Exaire JE, Sandoval J. Left main coronary artery extrinsic compression syndrome: a combined intravascular ultrasound and pressure wire. *J Invasive Cardiol*. 2006; 18(3):E102–4. [PubMed: 16495602]

21. de Jesus Perez VA, Haddad F, Vagelos RH, et al. Angina associated with left main coronary artery compression in pulmonary hypertension. *The Journal of heart and lung transplantation : the official publication of the International Society for Heart Transplantation*. 2009; 28(5):527–30. doi: 10.1016/j.healun.2008.12.008[published Online First: Epub Date]].
22. Caldera AE, Cruz-Gonzalez I, Bezerra HG, et al. Endovascular therapy for left main compression syndrome. Case report and literature review. *Chest*. 2009; 135(6):1648–50. doi: 10.1378/chest.08-2922[published Online First: Epub Date]]. [PubMed: 19497900]
23. Bugiardini R, Galvani M, Ferrini D, et al. Myocardial ischemia induced by prostacyclin and iloprost. *Clinical pharmacology and therapeutics*. 1985; 38(1):101–8. [PubMed: 2408809]
24. Heikkinen J, Milger K, Alexandre-Lafont E, et al. Cardiovascular Syndrome (Ortner's Syndrome) Associated with Chronic Thromboembolic Pulmonary Hypertension and Giant Pulmonary Artery Aneurysm: Case Report and Review of the Literature. *Case reports in medicine*. 2012; 2012:230736. doi: 10.1155/2012/230736[published Online First: Epub Date]]. [PubMed: 23424588]
25. Subramaniam V, Herle A, Mohammed N, et al. Ortner's syndrome: case series and literature review. *Brazilian journal of otorhinolaryngology*. 2011; 77(5):559–62. [PubMed: 22030961]
26. Nakao M, Sawayama T, Samukawa M, et al. Left recurrent laryngeal nerve palsy associated with primary pulmonary hypertension and patent ductus arteriosus. *Journal of the American College of Cardiology*. 1985; 5(3):788–92. [PubMed: 3973279]
27. Wilmschurst PT, Webb-Peploe MM, Corker RJ. Left recurrent laryngeal nerve palsy associated with primary pulmonary hypertension and recurrent pulmonary embolism. *British heart journal*. 1983; 49(2):141–3. [PubMed: 6824533]
28. Putta Rajasekhar PR, Bingi Prathap, Pani Chakra. A Rare Case of Primary Pulmonary Hypertension with Cardiovascular Syndrome. *Indian Journal of Applied Research*. 2014; 4(9):407–9.
29. Andjelkovic K, Kalimanovska-Ostic D, Djukic M, et al. Two rare conditions in an Eisenmenger patient: left main coronary artery compression and Ortner's syndrome due to pulmonary artery dilatation. *Heart & lung : the journal of critical care*. 2013; 42(5):382–6. doi: 10.1016/j.hrtlng.2013.06.001[published Online First: Epub Date]]. [PubMed: 23831302]
30. Achouh L, Montani D, Garcia G, et al. Pulmonary arterial hypertension masquerading as severe refractory asthma. *The European respiratory journal*. 2008; 32(2):513–6. doi: 10.1183/09031936.00005408[published Online First: Epub Date]]. [PubMed: 18669791]
31. Nakahira M, Nakatani H, Takeda T. Left vocal cord paralysis associated with long-standing patent ductus arteriosus. *AJNR American journal of neuroradiology*. 2001; 22(4):759–61. [PubMed: 11290495]
32. Albertini RE. Vocal cord paralysis associated with pulmonary emboli. *Chest*. 1972; 62(4):508–10. [PubMed: 5078011]
33. Camishion RC, Gibbon JH Jr, Pierucci L Jr. Paralysis of the left recurrent laryngeal nerve secondary to mitral valvular disease. Report of two cases and literature review. *Annals of surgery*. 1966; 163(6):818–28. [PubMed: 5933796]
34. Perloff JK, Hart EM, Greaves SM, et al. Proximal pulmonary arterial and intrapulmonary radiologic features of Eisenmenger syndrome and primary pulmonary hypertension. *The American journal of cardiology*. 2003; 92(2):182–7. [PubMed: 12860221]
35. Gupta V, Tonelli AR, Krasuski RA. Congenital heart disease and pulmonary hypertension. *Heart failure clinics*. 2012; 8(3):427–45. doi: 10.1016/j.hfc.2012.04.002[published Online First: Epub Date]]. [PubMed: 22748904]
36. Ditchfield MR, Culham JA. Assessment of airways compression by MR imaging in children with aneurysmal pulmonary arteries. *Pediatric radiology*. 1995; 25(3):190–1. [PubMed: 7644299]
37. Stanger P, Lucas RV Jr, Edwards JE. Anatomic factors causing respiratory distress in acyanotic congenital cardiac disease. Special reference to bronchial obstruction. *Pediatrics*. 1969; 43(5):760–9. [PubMed: 4238844]
38. Diaz-Guzman E, Vadi S, Minai OA, et al. Safety of diagnostic bronchoscopy in patients with pulmonary hypertension. *Respiration; international review of thoracic diseases*. 2009; 77(3):292–7. doi: 10.1159/000197465[published Online First: Epub Date]].

39. Maynar M, Lopez L, Gorriz E, et al. Massive brachiocephalic artery bleeding due to a Gianturco tracheal stent. *Journal of vascular and interventional radiology : JVIR*. 1993; 4(2):289–91. [PubMed: 8481580]
40. Stotz WH, Berkowitz ID, Hoehner JC, et al. Fatal complication from a balloon-expandable tracheal stent in a child: a case report. *Pediatric critical care medicine : a journal of the Society of Critical Care Medicine and the World Federation of Pediatric Intensive and Critical Care Societies*. 2003; 4(1):115–7. doi: 10.1097/01.PCC.0000043311.87364.7E[published Online First: Epub Date]].
41. Ramakrishna G, Sprung J, Ravi BS, et al. Impact of pulmonary hypertension on the outcomes of noncardiac surgery: predictors of perioperative morbidity and mortality. *Journal of the American College of Cardiology*. 2005; 45(10):1691–9. doi: 10.1016/j.jacc.2005.02.055[published Online First: Epub Date]]. [PubMed: 15893189]
42. Malm CJ, Ternstrom L, Jorgensen K, et al. Pulmonary artery dissection in a patient with undiagnosed pulmonary hypertension--A case report and review of literature. *Heart Lung*. 2015; 44(5):453–7. doi: 10.1016/j.hrtlng.2015.06.006[published Online First: Epub Date]]. [PubMed: 26153448]
43. Demerouti EA, Manginas AN, Athanassopoulos GD, et al. Complications leading to sudden cardiac death in pulmonary arterial hypertension. *Respir Care*. 2013; 58(7):1246–54. doi: 10.1181/respcare.02252[published Online First: Epub Date]]. [PubMed: 23271814]
44. Khattar RS, Fox DJ, Alty JE, et al. Pulmonary artery dissection: an emerging cardiovascular complication in surviving patients with chronic pulmonary hypertension. *Heart*. 2005; 91(2):142–5. doi: 10.1136/hrt.2004.045799[published Online First: Epub Date]]. [PubMed: 15657218]
45. Zylkowska J, Kurzyna M, Florkczyk M, et al. Pulmonary artery dilatation correlates with the risk of unexpected death in chronic arterial or thromboembolic pulmonary hypertension. *Chest*. 2012; 142(6):1406–16. doi: 10.1378/chest.11-2794[published Online First: Epub Date]]. [PubMed: 22797193]
46. Bijl M, Bronzwaer JG, van Rossum AC, et al. Angina pectoris due to left main coronary artery compression in Eisenmenger ductus arteriosus. *American heart journal*. 1993; 125(6):1767–71. [PubMed: 8498323]
47. Higgins JC, O'Brien JK, Battle RW, et al. Left main coronary artery compression in patent ductus arteriosus. *American heart journal*. 1993; 125(1):236–9. [PubMed: 8417525]
48. Kothari SS, Chatterjee SS, Sharma S, et al. Left main coronary artery compression by dilated main pulmonary artery in atrial septal defect. *Indian heart journal*. 1994; 46(4):165–7. [PubMed: 7875705]
49. Patrat JF, Jondeau G, Dubourg O, et al. Left main coronary artery compression during primary pulmonary hypertension. *Chest*. 1997; 112(3):842–3. [PubMed: 9315824]
50. Lee J, Kwon HM, Hong BK, et al. Total occlusion of left main coronary artery by dilated main pulmonary artery in a patient with severe pulmonary hypertension. *The Korean journal of internal medicine*. 2001; 16(4):265–9. [PubMed: 11855158]
51. Gullu H, Kosar F, Battaloglu B. Left main coronary artery compression by dilated pulmonary trunk in a patient with atrial septal defect. *Acta Cardiol*. 2003; 58(4):355–7. doi: 10.1021/AC.58.4.2005294[published Online First: Epub Date]]. [PubMed: 12948042]
52. Eksinar S, Gedevarishvili A, Koroglu M, et al. Extrinsic compression of the left main coronary artery in pulmonary hypertension. *JBR-BTR : organe de la Societe royale belge de radiologie*. 2005; 88(4):190–2.
53. Amaral FT, Alves L Jr, Granzotti JA, et al. Extrinsic compression of left main coronary artery from aneurysmal dilation of pulmonary trunk in a adolescent: involution after surgery occlusion of sinus venosus atrial septal defect and pulmonary trunk plasty for reduction. *Arquivos brasileiros de cardiologia*. 2007; 88(2):e40–3. [PubMed: 17384825]
54. Dodd JD, Maree A, Palacios I, et al. Images in cardiovascular medicine. Left main coronary artery compression syndrome: evaluation with 64-slice cardiac multidetector computed tomography. *Circulation*. 2007; 115(1):e7–8. doi: 10.1161/CIRCULATIONAHA.106.645622[published Online First: Epub Date]]. [PubMed: 17200448]
55. Dubois CL, Dymarkowski S, Van Cleemput J. Compression of the left main coronary artery by the pulmonary artery in a patient with the Eisenmenger syndrome. *European heart journal*. 2007;

28(16):1945. doi: 10.1093/eurheartj/ehl556[published Online First: Epub Date]]. [PubMed: 17311829]

56. Jo Y, Kawamura A, Jinzaki M, et al. Extrinsic compression of the left main coronary artery by atrial septal defect. The Annals of thoracic surgery. 2008; 86(6):1987–9. doi: 10.1016/j.athoracsur.2008.05.046[published Online First: Epub Date]]. [PubMed: 19022030]
57. Sivakumar K, Rajan M, Francis G, et al. Extrinsic compression of the left coronary ostium by the pulmonary trunk: management in a case of Eisenmenger syndrome. Texas Heart Institute journal / from the Texas Heart Institute of St Luke's Episcopal Hospital, Texas Children's Hospital. 2010; 37(1):95–8.
58. Vaseghi M, Lee MS, Currier J, et al. Percutaneous intervention of left main coronary artery compression by pulmonary artery aneurysm. Catheterization and cardiovascular interventions : official journal of the Society for Cardiac Angiography & Interventions. 2010; 76(3):352–6. doi: 10.1002/ccd.22555[published Online First: Epub Date]]. [PubMed: 20552652]
59. Qian J, Chen Z, Zhou D, et al. Compression of left main coronary artery by a dilated pulmonary artery in a female patient with atrial septal defect. International journal of cardiology. 2012; 161(2):e23–5. doi: 10.1016/j.ijcard.2012.03.130[published Online First: Epub Date]]. [PubMed: 22483622]
60. Lee MS, Mahmud E. Percutaneous revascularization for left main coronary artery compression from pulmonary artery enlargement due to pulmonary hypertension. Reviews in cardiovascular medicine. 2012; 13(1):e32–6. [PubMed: 22565536]
61. Doyen D, Moceri P, Moschietto S, et al. Left main coronary artery compression associated with primary pulmonary hypertension. Journal of the American College of Cardiology. 2012; 60(6): 559. doi: 10.1016/j.jacc.2011.12.059[published Online First: Epub Date]]. [PubMed: 22858290]
62. Sahay S, Tonelli AR. Ventricular fibrillation caused by extrinsic compression of the left main coronary artery. Heart. 2013; 99(12):895–6. doi: 10.1136/heartjnl-2012-303408[published Online First: Epub Date]]. [PubMed: 23286969]
63. Brinton WD. Primary pulmonary hypertension. British heart journal. 1950; 12(3):305–11. [PubMed: 15426692]
64. Soothill JF. A case of primary pulmonary hypertension with paralysed left vocal cord. Guy's Hospital reports. 1951; 100(3):232–7.
65. Shah KD, Shah UK. Hoarseness--a presenting manifestation of primary pulmonary hypertension. Indian heart journal. 1975; 27(2):105. [PubMed: 1184052]
66. Kagal AE, Shenoy PN, Nair KG. Ortner's syndrome associated with primary pulmonary hypertension. Journal of postgraduate medicine. 1975; 21(2):91–5. [PubMed: 126321]
67. Aszkenasy OM, Clarke TJ, Hickling P, et al. Systemic lupus erythematosus, pulmonary hypertension, and left recurrent laryngeal nerve palsy. Annals of the rheumatic diseases. 1987; 46(3):246–7. [PubMed: 3579390]
68. Soliman MS. Hoarseness in schistosomiasis. Chest. 1997; 112(4):1150. [PubMed: 9377941]
69. Sengupta A, Dubey SP, Chaudhuri D, et al. Ortner's syndrome revisited. The Journal of laryngology and otology. 1998; 112(4):377–9. [PubMed: 9659503]
70. Arimura FE, Santiago RRSS, Gavilanes F, et al. Idiopathic Pulmonary Arterial Hypertension Leading To Dynamic Bronchial Compression And Orthopnea: A Case Report Of The Use Of Electrical Impedance Tomography (eit) As A New Investigation Tool In Pulmonary Function Test. Am J Respir Crit Care Med. 2015; 191:A4909.

Figure 1. Left main coronary artery compression

Panel A: Posterior-anterior chest radiography showing bilaterally enlarged pulmonary arteries. Panel B: Computed tomography angiography of the aorta revealing aneurysmal dilation of the main pulmonary artery compressing the LMCA (white arrow). Panel C: Computed tomography showing a stent in the LMCA (white arrow). Panel D and E: Coronary angiography with proximal narrowing of the LMCA before stent placement (panel D, black arrow) and after stent implantation (panel E, black arrow). Insert panel: intravascular ultrasound of the LMCA showing a severely reduced intraluminal area at 5 mm². Panel F: Computed tomography showing compression of the proximal portion of the main bronchi (black arrow).

Figure 2. Left recurrent laryngeal nerve compression

Computed tomography of the neck and chest with contrast showing a fold on the left vocal cord suggestive of palsy (white arrow in Panel A). Rest of the panels reveal axial cuts at different levels including aortic arch (panel B), aorto-pulmonary window (panel C) and pulmonary artery bifurcation (panel D).

Figure 3. Tracheobronchial tree compression

Panel A: Posterior-anterior chest X ray showing dextrocardia, cardiomegaly and severely dilated pulmonary arteries. The convex bulging opacity over the right mediastinum represents an enlarged right pulmonary artery causing compression of the right main stem bronchus (black arrow). Panel B and C: Computed tomography of the chest without contrast revealing narrowing of the right main bronchus (white arrow in Panel B) with distal bronchiectasis (Panel C). Panel D to F depict magnetic resonance images of the chest, demonstrating severely dilated pulmonary arteries. Abbreviations: Ao: aorta, LPA: left pulmonary artery, RPA: right pulmonary artery.

Figure 4. Compression of adjacent anatomical structures by a dilated pulmonary artery
Panel A: represents coronal section illustrating a dilated main pulmonary artery with compression of the LRLN between the pulmonary artery and the aortic arch, and of the LMCA at its origin. **Panel B:** illustrates a severe enlargement of the main pulmonary artery with compression of both main bronchi. Figure used by permission of the Cleveland Clinic.

Table 1
Reports of left main coronary artery compression by enlarged pulmonary artery in adults (16 years old) with PAH.

Study, year, ref	Ref	n	Age (years) [*]	Gender	Type of PAH	PAH-specific therapy	Time from diagnosis or symptoms to complication [*] (years)	LMCA Stenosis (%) [*]	Size of PA (cm) [*]	PA/Ao ratio [*]	mPAP (mmHg) [*]	Treatment	Symptoms improved (yes/no)
Fujiwara et al 1992	[17]	3	45,64,58	1M,2F	ASD	No	N/A	50,50,75	N/A	N/A	N/A	3 ASD closure,1 CABG	Yes
Bijl et al 1993	[46]	1	45	M	PDA	Yes	N/A	N/A	N/A	N/A	100	No intervention	No
Higgins et al 1993	[47]	1	38	F	PDA	No	1	>50	N/A	N/A	127	Heart-lung transplant	No
Kothari et al 1994	[48]	2	43,43	M,F	ASD	No	N/A	>50	N/A	N/A	N/A,30	ASD closure	No
Patrat et al 1997	[49]	1	37	F	IPAH	No	N/A	>50	N/A	N/A	90	Heart-lung transplant	Yes
Kawut et al 1999	[5]	3	45,55,57	1F,2M	ASD,VSD,1PAH	No	6,27,26	80,80,80	6.4,6.7,12	N/A	75,53,50	N/A	No
Kajita et al 2001	[18]	8	17-55	7F,1M	4 ASD, 1 TOF & APV, 1 PDA & VSD, 1 schistosomiasis mansonii, 1 IPAH	No	N/A	50-85	3.9-6.5	1.7-2.6	38-88	3 CABG	No
Rich et al 2001	[14]	2	71,53	F	IPAH	Yes	12,2	90,85	N/A	N/A	70,51	Stent	Yes
Lee at al 2001	[50]	1	34	F	ASD	No	N/A	>50	4.0	N/A	N/A	ASD closure	No
Gullu et al 2003	[51]	1	52	M	ASD	No	0.5	90	N/A	N/A	41	ASD closure, CABG	Yes
Gomez Varela et al 2004	[7]	1	31	F	IPAH	Yes	3	80	4.0	N/A	49	Stent	Yes
Mesquita et al 2004	[13]	7	26-58	6F,1M	5 ASD, 1IPAH, 1 AVSD	No	N/A	50-90	4.0-7.3	1.2-2.6	34-64	1 pulmonary trunk reduction, 3 CABG	Yes
Eksinar et al 2005	[52]	1	51	F	ASD	No	3	80	N/A	N/A	42	ASD closure	Yes
Piña et al 2006	[20]	1	55	F	ASD	No	N/A	70	N/A	N/A	74	No intervention as IVUS showed mild stenosis,	No
Vaseghi et al 2007	[8]	1	51	F	Rheumatoid arthritis	Yes	15	70	11.0	N/A	39	Stent	Yes
Amaral et al 2007	[53]	1	16	F	ASD with APVR	No	N/A	>50	N/A	N/A	83	ASD closure and redirection of the APVR	Yes
Dodd et al 2007	[54]	1	28	M	PDA	No	N/A	>90	6.0	2.3	N/A	Stent	Yes
Dubois et al 2007	[55]	1	51	F	ASD	No	N/A	>50	N/A	N/A	N/A	Stent	Yes
Lindsey et al 2008	[9]	1	53	F	Drug induced PAH	Yes	N/A	90	4.4	N/A	71	Stent	Yes

Study, year, ref	Ref	n	Age (years) *	Gender	Type of PAH	PAH-specific therapy	Time from diagnosis or symptoms to complication *	LMCA Stenosis (%) *	Size of PA * (cm)	PA/Ao ratio *	mPAP (mmHg) *	Treatment	Symptoms improved (yes/no)
Jo et al 2008	[56]	1	42	F	ASD	No	N/A	75	4.7	N/A	40	ASD closure, CABG	Yes
De Jesus Perez et al 2009	[21]	2	58,31	M	CREST syndrome, drug induced PAH	Yes	N/A	50,75	3.5	N/A	60	Stent	Yes
Caldera et al 2009	[22]	1	48	F	PDA	Yes	4	90	6.4	1.89	90	Stent	Yes
Sivakumar et al 2010	[57]	1	58	M	PDA	No	N/A	N/A	N/A	N/A	N/A	Stent	Yes
Vaseghi et al 2010	[58]	5	28-67	F	3 IPAH, 2 ASD	4 yes, 1 no	1-14	60-95	4.2-11.0	N/A	N/A	Stent	Yes
Qian et al 2012	[59]	1	19	F	ASD	No	N/A	75	4.3	N/A	68	Stent	Yes
Lee et al 2012	[60]	1	46	F	ASD	No	N/A	N/A	N/A	N/A	N/A	Stent	Yes
Demkow et al 2012	[19]	1	29	F	IPAH	Yes	N/A	90	7	N/A	N/A	Stent	Yes
Doyen et al 2012	[61]	1	61	M	IPAH	No	N/A	N/A	N/A	N/A	N/A	Stent	Yes
Andjelkovic et al 2013	[29]	1	37	F	ASD	Yes	N/A	90	4.3	1.77	76	Stent	Yes
Sahay et al 2013	[62]	1	43	F	VSD	Yes	N/A	80	N/A	N/A	N/A	Stent	Yes

Abbreviations: APV: absent pulmonary valve, APVR: anomalous pulmonary venous return, ASD: atrial septal defect, CABG: coronary artery bypass graft, F: female, IPAH: idiopathic pulmonary arterial hypertension, IVUS: intravascular ultrasound, LMCA: left main coronary artery, M: male, mPAP: mean pulmonary arterial pressure, N/A: not available, PA: pulmonary artery, PAH: pulmonary arterial hypertension, PDA: patent ductus arteriosus, Ref: reference, TOF: tetralogy of Fallot, VSD: ventricular septal defect.

*When more than 3 patients, data are given as range.

Table 2

Reports of left recurrent laryngeal nerve compression by enlarged pulmonary artery in adults with PAH.

Study, year, ref	Ref	n	Age (years)	Gender	Type of PAH	PAH-specific therapy	Hoarseness at presentation (yes/no)	Time from SOB and development of hoarseness (months)	Size of PA (cm)	mPAP (mmHg)	Improvement of hoarseness (Yes/no)
Brinton et al 1950	[63]	1	26	M	IPAH	No	No	N/A	N/A	N/A	No
Soothill et al 1951	[64]	1	22	M	IPAH	No	Yes	9	N/A	98	No
Shah et al 1975	[65]	10	N/A	N/A	IPAH	No	Yes in 6	N/A	N/A	N/A	No
Kagal et al 1975	[66]	2	28,25	1M,1F	IPAH	No	Yes in 1	2,4	N/A	N/A	No
Wilmshurst et al 1983	[27]	1	37	M	IPAH	Yes	Yes	4	N/A	57	No
Nakao et al 1985	[26]	2	34,57	F	IPAH, PDA	No	Yes	N/A	N/A	50,52	No
Aszkenasy et al 1987	[67]	1	41	F	SLE induced PAH	Yes	No	1	N/A	77	Yes
Soliman 1997	[68]	1	27	M	Schistosomiasis associated PAH	No	Yes	N/A	N/A	49	No
Sengupta et al 1998	[69]	1	37	M	IPAH	No	Yes	8	2.3	62	No
Nakahira et al 2001	[31]	1	76	F	PDA	No	Yes	Simultaneously	N/A	29	No
Achouh et al 2008	[30]	1	56	M	ASD	Yes	No	60	N/A	60	No
Andjelkovic et al 2013	[29]	1	37	F	ASD	Yes	No	Simultaneously	43	76	No
Shankar et al 2014	[3]	1	19	M	IPAH	Yes	Yes	11	3.8	101	Yes
Rajasekhar et al 2014	[28]	1	35	F	IPAH	Yes	Yes	Simultaneously	N/A	N/A	Yes

Abbreviations: ASD: atrial septal defect, F: female, IPAH: idiopathic pulmonary arterial hypertension, M: male, mPAP: mean pulmonary arterial pressure, N/A: not available, PA: pulmonary artery, PAH: pulmonary arterial hypertension, PDA: patent ductus arteriosus, Ref: reference, SLE: systemic lupus erythematosus.

Table 3
Reports of tracheobronchial compression by enlarged pulmonary artery in adults (16 years old) with PAH.

Study, year, ref	Ref	n	Age (years)	Gender	Type of PAH	PAH-specific therapy	Site of compression	Size of PA (cm)
Edwards et al 1960	[4]	2	24,N/A	M,N/A	ASD,PDA	No	left tracheal border, LB, RMB, LUB	N/A
Perloff et al 2003	[34]	1	54	M	VSD	No	RMB	N/A
Achouh et al 2008	[30]	2	34,56	2 M	Mesenterico-caval shunt, ASD	Yes	LB,2 RB	N/A
Arimura et al 2015	[70]	1	42	F	I PAH	Yes	LB	7.9

Abbreviations: ASD: atrial septal defect, IPAH: idiopathic pulmonary arterial hypertension, LB: left main stem bronchus, LUB: left upper lobe bronchus, N/A: not available, PA: pulmonary artery, PDA: patent ductus arteriosus, PHA: pulmonary arterial hypertension, Ref: reference, RB: right main stem bronchus, RMB: right middle bronchus, VSD: ventricular septal defect.