PULMONARY VASCULAR DISEASES. pdf

1: Vascular Disease Overview | Cleveland Clinic

Pulmonary vascular disease is the medical term for disease affecting the blood vessels leading to or from the lungs. Most forms of pulmonary vascular disease cause shortness of breath.

Conditions like a pulmonary embolism can occur more rapidly and lead to death sooner than pulmonary hypertension, which will have a gradual onset of symptoms. And if your child has other serious conditions and begins to demonstrate symptoms of PVD, seek immediate medical assistance. PVD is often caused by some other serious condition, including: Lung disease Blood clot elsewhere in the body Failure to absorb a blood clot Congestive [kuhn-JEST-iv] heart failure Damaged heart valves There are some genetic risk factors for PVD and some research indicates it can be more common at higher altitudes and for children who are overweight. Treatments There are two primary methods of treatment for pulmonary vascular disease as listed below. Blood thinners or blood pressure medicine may be prescribed to control the pulmonary hypertension. Surgery may be needed to repair damaged arteries. However, some simple lifestyle changes that can help avoid risk factors for pulmonary vascular disease, including: What are Pulmonary Vascular Diseases? The blood vessels between the lungs and heart they capture oxygen in the lungs and to be used by the rest of the body. If this process is slowed or stopped, carbon dioxide can build up on the body, which can cause a number of problems. Pulmonary arterial [ahr-TEER-ee-uh l] hypertension. This condition involves increased blood pressure in the arteries carrying blood to the lungs. Pulmonary venous [VEE-nuh s] hypertension. This condition is increased blood pressure in the veins carrying blood away from the heart. A blood clot, usually from a deep vein in the leg, travels to the heart and is pumped into the lungs. This can also lead to a condition called chronic thromboembolic [throm-boh-EM-buh-lik] disease in which a blood clot is not absorbed completely in the lungs and causes other vessels around it to become diseased. Pulmonary vascular disease is sometimes referred to as PVD, but should not be confused with peripheral [puh-RIF-er-uh l] vascular disease, which is a narrowing of the vessels supplying blood to the legs, arms, stomach, or kidneys. The content presented here is for your information only. It is not a substitute for professional medical advice, and it should not be used to diagnose or treat a health problem or disease. Please consult your healthcare provider if you have any questions or concerns. Healthcare Intermountain Healthcare is a Utah-based, not-for-profit system of 23 hospitals, a Medical Group with more than 1, physicians and advanced practice clinicians at about clinics, a health plans division called SelectHealth, and other health services. Helping people live the healthiest lives possible, Intermountain is widely recognized as a leader in clinical quality improvement and in efficient healthcare delivery.

PULMONARY VASCULAR DISEASES. pdf

2: Pulmonary Vascular Diseases

Pulmonary Vascular Disease in simple form can be defined as any medical condition that affects the blood vessels between the heart and lungs. The process of oxygenation of blood takes place in the following way.

Back to Diseases and Conditions Pulmonary Vascular Disease Pulmonary vascular disease PVD is a broad term including any condition that affects the blood vessels within the lungs. These vessels take blood that is depleted of oxygen to the lungs from the right side of the heart. Deoxygenated blood travels through the pulmonary arteries where oxygen is taken up. The pulmonary veins leave the lungs and take blood rich with oxygen to the left side of the heart where oxygenated blood is distributed throughout the body. This process continually replenishes the blood with oxygen, and lets carbon dioxide be exhaled. Our pulmonologists and cardiologists work closely together. This expertise and collaboration helps patients manage symptoms and have an improved quality of life. The research on PVD which is conducted at BWH provides greater understanding of these diseases and is translated directly into exceptional patient care. It occurs when the small blood vessels that go through the lungs become thicker, constrict tighter or become plugged. This leads to increased pressure in those vessels making the heart work harder to pump blood. If high pressure remains, the heart will become enlarged and weaker, pumping blood less efficiently into the lungs and eventually throughout the body. Types of PH include: Pulmonary Arterial Hypertension Increased blood pressure in the pulmonary arteries carrying blood away from the right side of the heart to the lungs. Pulmonary arterial hypertension can be caused by heart failure, lung disease or an autoimmune disease. If there is no apparent cause, it is called idiopathic pulmonary arterial hypertension. Pulmonary Venous Hypertension Increased blood pressure in the pulmonary veins carrying blood away from the lungs to the left side of the heart. Pulmonary venous hypertension is usually caused by poorly controlled systemic hypertension and congestive heart failure. A damaged mitral or aortic valve in the heart mitral or aortic valve stenosis or mitral regurgitation may also contribute to pulmonary venous hypertension. Our pulmonary vascular team has experience diagnosing CTEPH and in both the medical and surgical treatment of this condition. Some, but not all, patients have a history of blood clots to the lung. Correct diagnosis is important because in many cases a surgical procedure called pulmonary thromboendarterectomy can successfully treat this condition. This procedure may be recommended for people whose pulmonary hypertension is due to persistent pulmonary emboli blood clots. Symptoms of Pulmonary Hypertension.

3: What is Pulmonary Hypertension (PH) - Brigham and Women's Hospital

Pulmonary vascular disease is sometimes referred to as PVD, but should not be confused with peripheral [puh-RIF-er-uh l] vascular disease, which is a narrowing of the vessels supplying blood to the legs, arms, stomach, or kidneys.

As the heart beats, it pumps blood through a system of blood vessels, called the circulatory system. The vessels are elastic tubes that carry blood to every part of the body Arteries carry blood away from the heart. Veins return blood back to the heart. Vascular Disease includes any condition that affects your circulatory system, such as peripheral artery disease. This ranges from diseases of your arteries, veins and lymph vessels to blood disorders that affect circulation. The following are conditions that fall under the category of "Vascular Disease": Arterial Disease Peripheral Artery Disease Like the blood vessels of the heart coronary arteries , your peripheral arteries blood vessels outside your heart also may develop atherosclerosis, the build-up of fat and cholesterol deposits, called plaque, on the inside walls. Over time, the build-up narrows the artery. Eventually the narrowed artery causes less blood to flow, and a condition called ischemia can occur. Types of peripheral arterial disease include: A blockage in the legs can lead to leg pain or cramps with activity claudication , changes in skin color, sores or ulcers and feeling tired in the legs. Total loss of circulation can lead to gangrene and loss of a limb. A blockage in the blood vessels leading to the gastrointestinal system Renal artery disease: A blockage in the renal arteries can cause renal artery disease stenosis. The symptoms include uncontrolled hypertension high blood pressure , congestive heart failure, and abnormal kidney function. The muscle and tendons near the knee compress the popliteal artery, restricting blood flow to the lower leg and possibly damaging the artery. Although the cause is unknown, there is a strong association with tobacco use or exposure. The arteries of the arms and legs become narrowed or blocked, causing lack of blood supply ischemia to the fingers, hands, toes and feet. Pain occurs in the arms, hands, and more frequently the legs and feet, even at rest. With severe blockages, the tissue may die gangrene , requiring amputation of the fingers and toes. Carotid Artery Disease Carotid artery disease is a blockage or narrowing in the arteries supplying the brain, and can lead to a transient ischemic attack TIA or stroke Carotid artery dissection begins as a tear in one layer of the artery wall. Blood leaks through this tear and spreads between the layers of the wall. Carotid body tumors are growths within the nervous tissue around the carotid artery Carotid artery aneurysm Venous Disease Veins are flexible, hollow tubes with flaps inside, called valves. When your muscles contract, the valves open, and blood moves through the veins. When your muscles relax, the valves close, keeping blood flowing in one direction through the veins. If the valves inside your veins become damaged, the valves may not close completely. This allows blood to flow in both directions. When your muscles relax, the valves inside the damaged vein s will not be able to hold the blood. This can cause pooling of blood or swelling in the veins. The veins bulge and appear as ropes under the skin. The blood begins to move more slowly through the veins, it may stick to the sides of the vessel walls and blood clots can form. Varicose veins are bulging, swollen, purple, ropy veins, seen just under your skin, caused by damaged valves within the veins. Spider veins are small red or purple bursts on your knees, calves, or thighs, caused by swollen capillaries small blood vessels Klippel-Trenaunay syndrome KTS , a rare congenital vascular disorder May-Thurner syndrome MTS is caused when the left iliac vein is compressed by the right iliac artery, which increases the risk of deep vein thrombosis DVT in the left extremity. Blood Clots A clot forms when clotting factors in the blood cause it to coagulate or become a solid, jelly-like mass. When a blood clot forms inside a blood vessel a thrombus , it can dislodge and travel through the blood stream, causing a deep vein thrombosis, pulmonary embolis, heart attack or stroke. Blood clots in the arteries can increase the risk for stroke, heart attack, severe leg pain, difficulty walking, or even the loss of a limb. Hypercoagulable states are conditions that put people at increased risk for developing blood clots. Deep vein thrombosis DVT is a blood clot occurring in a deep vein. Pulmonary embolism is a blood clot that breaks loose from a vein and travels to the lungs. Axillo-subclavian vein thrombosis , also called Paget-Schroetter Syndrome, is a most common vascular conditions to affect young, competitive athletes. The condition develops when a vein in the armpit the axilla or

in the front of the shoulder the subclavian vein is compressed by the collarbone clavicle , the first rib, or the surrounding muscle, increasing risk for blood clots. Superficial thrombophlebitis is a blood clot in a vein just under the skin Aortic Aneurysm An aneurysm is an abnormal bulge in the wall of a blood vessel. Aneurysms can form in any blood vessel, but they occur most commonly in the aorta aortic aneurysm which is the main blood vessel leaving the heart: Thoracic aortic aneurysm part of aorta in the chest Abdominal aortic aneurysm - include one or more of the following: Patients with FMD have abnormal cellular growth in the walls of their medium and large arteries. This can cause the arteries with the abnormal growth to look beaded. The arteries may also become narrow stenosis. Other vascular conditions include: Blood clotting disorders are disorders that make the blood more likely to form blood clots hypercoagulable in the arteries and veins. These conditions may be inherited congenital, occurring at birth or acquired during life and include: Elevated levels of factors in the blood which cause blood to clot fibrinogen, factor 8, prothrombin Deficiency of natural anticoagulant blood-thinning proteins antithrombin, protein C, protein S Elevated blood counts Abnormal Fibrinolysis the breakdown of fibrin Abnormal changes in the lining of the blood vessels endothelium Lymphedema The lymphatic system is a circulatory system that includes an extensive network of lymph vessels and lymph nodes. Lymphedema is an abnormal build-up of fluid that causes swelling, most often in the arms or legs. Lymphedema develops when lymph vessels or lymph nodes are missing, impaired, damaged or removed. Primary lymphedema is rare and is caused by the absence of certain lymph vessels at birth, or it may be caused by abnormalities in the lymphatic vessels. Secondary lymphedema occurs as a result of a blockage or interruption that alters the lymphatic system. Secondary lymphedema can develop from an infection, malignancy, surgery, scar tissue formation, trauma, deep vein thrombosis DVT , radiation or other cancer treatment. Doctors vary in quality due to differences in training and experience; hospitals differ in the number of services available. The more complex your medical problem, the greater these differences in quality become and the more they matter.

PULMONARY VASCULAR DISEASES. pdf

4: Pulmonary Vascular Disease Overview - Brigham and Women's Hospital

Like pulmonary arterial hypertension, this disease can be caused by lung disease, autoimmune disease or in some cases heart failure. If there is a damaged valve within the heart, this can cause the pulmonary venous hypertension to occur.

The law requires that our healthcare facilities and medical personnel protect the privacy of your medical record and other health information We are also required to notify you of our privacy practices with regard to your Protected Health Information. The Lung Institute collects information by various methods including information actively provided by its lead providers, customers and information arising from customer surveys and general feedback. The types of personal information we collect include name, contact information, identification information, credit information and other data types as appropriate. Credit card information is used for billing purposes only. How We Use This Information: This information is used to aid in the provision of our various products and services, including customer service, accounting, billing, collections and the marketing of other products and services. The Lung Institute may use aggregate or anonymous information for various uses for itself and third parties. The Lung Institute does not share personal information with any third parties except as disclosed in this policy. Security Personal information is stored in a combination of paper and electronic files. They are protected by security measures appropriate to the nature of the information. If an individual believes that any of their personal information is inaccurate, we will make appropriate corrections. Cookies are used by the Lung Institute for the convenience of our users. Cookies automatically authenticate the user. However, in doing so they may find themselves challenged for username and password information on multiple occasions. When you click on one of these links, you are moving to another website. We encourage you to read the privacy statements of these linked sites as their privacy policy may differ from ours. Telephone Numbers and Calls: By providing your telephone number to the Lung Institute on this contact form, you acknowledge and agree that you may receive a telephone call from the Lung Institute, at the number you provided on the online form, to receive information regarding the treatments offered at the Lung Institute. By providing your mobile number to the Lung Institute, you further acknowledge and agree that you may receive text messages SMS from the Lung Institute at the number you provided to the Lung Institute. Protected Health Information will not be requested by the Lung Institute, if you choose to respond with Protected Health Information you agree to release the Lung Institute from any liability related to such disclosure. The Lung Institute may amend this policy from time to time. If such amendments affect how the Lung Institute uses or discloses personal information already held by the Lung Institute in a material way, the Lung Institute will obtain consent. Notwithstanding the general terms of this policy, the collection, use, and disclosure of personal information may be made outside of the terms herein to the extent provided for in any applicable privacy or other legislation in effect from time to time.

PULMONARY VASCULAR DISEASES. pdf

5: Pulmonary Vascular Disease | Pulmonary, Critical Care and Sleep Medicine

Pulmonary vascular disease (PVD) is a broad term including any condition that affects the blood vessels within the lungs. These vessels take blood that is depleted of oxygen to the lungs from the right side of the heart.

Pulmonary vascular disease is a term for a disease affecting the blood vessels leading to or from the lungs. It refers to a category of disorders. Below is some basic information about the major types of pulmonary vascular disease. Symptoms include difficulty breathing, chest pain, fainting, and a rapid heart rate. A pulmonary embolism can damage the heart or cause death. It can be prevented by drugs that break up blood clots, proper exercise and physical activity, compression socks, and pneumatic compression. It often progresses gradually, causing small blood vessels in the lungs to become diseased until a large part of the pulmonary arterial system is affected. Pulmonary veno-occlusive disease is a rare form of high blood pressure in the lung. It can be caused by a viral infection or as a complication of lupus, leukemia, lymphoma, chemotherapy, or bone marrow transplantation. However, in most cases, the cause is unknown. Symptoms include difficulty breathing, fatigue, fainting, and coughing up blood. Pulmonary veno-occlusive disease can cause narrowed pulmonary veins, pulmonary hypertension , congestion, and swelling of the lungs. There is no known cure, but medicine can be used to treat the disease. Arteriovenous malformations AVMS are tangles in the circulatory system that are usually present from birth. The cause of these malformations is unknown. Because the circulatory system carries blood between the heart and the rest of the blood, these tangles can interfere with the blood circulation in an organ. They generally occur in the head but can also be present in internal organs, the limbs, and the torso. The greatest danger of an AVM is hemorrhage, internal bleeding caused by a broken blood vessel. Treatment can include surgery or irradiation therapy. Pulmonary edema occurs when fluid builds up in the air sacs of the lungs. It is often caused by heart failure. When the heart fails, the blood pressure increases in the veins going through the lungs, causing fluid to be pushed into the lungs and resulting in a shortness of breath. Pulmonary edema can be caused by exercise at very high altitudes, damage to the lung by poisonous gas or sever infection, as a side effect of medication, or from major trauma. It can also be a complication of heart disease. Information on this page is for reference and educational purposes only. For more information about pulmonary vascular disease, talk to your doctor or primary care provider. He is a writer and designer. He is extensively trained on oxygen therapy products from leading manufacturers such as Inogen, Respironics, Chart, Invacare, ResMed and more. Scott works closely with respiratory therapists and oxygen specialists to educate the community about oxygen therapy products, COPD, asthma and lung diseases. He writes weekly columns and is passionate about educating the community on oxygen therapy and respiratory issues.

6: American Thoracic Society - Pulmonary Vascular Disease

Pulmonary vascular disease may lead to complications such as abnormal enlargement of the right side of the heart (cor pulmonale), irregular heartbeat (arrhythmia), bleeding, and pulmonary embolism (PE).

Causes Pulmonary embolism Pulmonary embolism is usually the consequence of a blood clot in the legs or pelvis. This blood clot can break off and travel along the venous system to the pulmonary arteries. Certain groups of people are more at risk of developing blood clots, including the elderly, people who have undergone medical or surgical events that have resulted in long periods of time in bed, people with a previous history of blood clots and those under hormone replacement therapy and oral contraceptive therapy. There are many different forms of pulmonary hypertension and each is treated very differently. Doctors class the disease into five main groups: Pulmonary arterial hypertension PAH due to different causes Pulmonary hypertension due to left heart diseases Pulmonary hypertension due to lung diseases or lack of oxygen hypoxia Chronic thromboembolic pulmonary hypertension, where the blood vessels are blocked or narrowed by blood clots Pulmonary hypertension with an unclear cause or several different triggers Prevention Pulmonary embolism Preventative medicines, such as heparin, can be given to reduce the chance of a blood clot and therefore, pulmonary embolism. Anti-vitamin K drugs can be given after a pulmonary embolism to reduce the risk of blood clotting. Oral anticoagulation drugs, that can be easily prescribed and require less monitoring, are currently in development. Pulmonary hypertension People with long-term lung diseases should be treated with oxygen therapy to prevent the development of pulmonary hypertension. There is no known method of preventing pulmonary arterial hypertension. Treatment Pulmonary embolism Pulmonary embolism is often difficult to diagnose. Tests to check for the condition also include a blood test, called a D-dimer, to rule out pulmonary embolism and computed tomography CT angiography, an imaging technique which uses x-rays, a CT scan and magnetic resonance imaging MRI. In an emergency situation, a test called a bedside echocardiography, which uses ultrasound to create images of the heart, can help to diagnose PE. Drugs known as anticoagulants can be used to treat pulmonary embolism. Supplementary oxygen can also be provided for people who are struggling to breathe. Thrombolytic drugs, which can dissolve the clots in the pulmonary arteries, are recommended for people who have persistent heart failure and a high risk of PE. Pulmonary hypertension Early diagnosis is difficult with pulmonary arterial hypertension PAH as many patients have no or few symptoms or just appear unfit. Many people go to the doctor late in the course of the disease with signs of right heart failure. When it is suspected two procedures can be used to help diagnose pulmonary hypertension. The first, known as an echocardiogram, uses sound waves to create a moving picture of the heart. The second, known as right heart catheterisation, is an invasive procedure where a thin tube, known as a catheter, is inserted into the right heart to measure how well it is pumping and into the pulmonary arteries to measure the blood pressure. Screening programmes that help to identify people most at risk of pulmonary hypertension are a useful way of diagnosing people early. There is no known cure for pulmonary hypertension but basic drugs such as anticoagulants or oxygen supplementation can help. Patients with pulmonary arterial hypertension can be treated with specific therapies, known as prostacyclins, endothelin receptor antagonists or type 5 phosphodiesterase inhibitors. If pulmonary hypertension is due to chronic thrombo-embolic disease, a surgical operation known as pulmonary endarterectomy can cure the disease by clearing the clot and scar material in the blood vessels arteries of the lungs. If the condition is severe, lung transplantation may be an option. If pulmonary hypertension is linked to long-term heart or lung diseases, treatment of the underlying condition is recommended. Data from World Health Organization World and Europe Mortality Databases, November update Current and Future Needs There is a need to develop better diagnostic methods for pulmonary embolism Antithrombotic prophylaxis with low-molecular weight herparin significantly reduces the risk of venous thromboembolic diseases in patients who are at risk, and thereby the risk of pulmonary embolism Better awareness of pulmonary hypertension is essential for the earlier diagnosis and management of the condition New therapies are needed for pulmonary arterial hypertension as there is no known cure Prevention methods should be improved for people at risk of pulmonary hypertension More research is needed

to determine the causes of pulmonary hypertension.

PULMONARY VASCULAR DISEASES. pdf

7: Pulmonary Complications of Collagen Vascular Diseases - The Clinical Advisor

Pulmonary vascular disease There are two main types of pulmonary vascular diseases: pulmonary embolism and pulmonary hypertension. Pulmonary embolism occurs due to blood clots which block branches of the arteries in the lungs, often following thrombosis in the veins of the leg or elsewhere.

Respiratory Problems Caused by Vascular Disease In case of pulmonary vascular disease all the symptoms and signs develop as a consequence of damage to the blood vessels that are located between the heart and the lungs. The blood delivered to the lungs is rich in carbon dioxide and after this gas is released and the blood enriched with oxygen it leaves the lungs and re-enters the heart, ready to be distributed across the entire body. Any damage to blood vessels that are in charge with transfer of blood from the heart to the lungs and back to the heart once again is a cause of pulmonary vascular disease. There are several condition that belong to this group of illnesses. They include pulmonary arterial hypertension, pulmonary venous hypertension, pulmonary embolism and chronic thromboembolic disease. Symptoms and signs patients suffering from pulmonary vascular disease experience basically depend on the type of damaged blood vessels, disease progression and the percent of the vascular system affected by the disease. Pulmonary Embolism is One of the most Common Issues In patients suffering from pulmonary embolism there is a blood clot or multiple blood clots blocking blood vessels in the lungs. These form somewhere else in the body and travel to the lungs. Large blood clots cause obstruction of blood vessels with large diameter such as the main artery of the lungs while smaller ones easily pass this blood vessel blocking smaller blood vessels and causing less severe form of the disease. In the majority of cases blood clots that trigger blockage originate from deep veins of the lower extremities. Furthermore, it is also possible to develop pulmonary embolism due to embolisation of air, fat, amniotic fluid or talc in drugs injected by intravenous drug users. Proximal leg deep venous thrombosis, however, remains the leading cause of pulmonary embolism. Changes in blood flow, changes in the vessel wall as well as factors affecting the quality of circulating blood determine whether the blood clot will develop in the first place and if it will eventually dislodge and migrate to other parts of the circulatory systems. The risk of venous thrombosis and subsequent pulmonary embolism is higher in people who are immobilized for a long period of time especially after surgery, injury etc. Increased susceptibility to blood formation is also reported to occur in women using estrogen-containing contraception and patients suffering from genetic or acquired thrombophilia. When it comes to symptoms and signs of pulmonary embolism, these include sudden onset of shortness of breath, rapid breathing and chest pain, the one that tends to become much worse when a person takes deep breaths. Cyanosis affects patients in whom the lungs do not receive sufficient amount of oxygen and cannot release excess of carbon dioxide. Although auscultation of the lungs may be normal, sometimes there may be a pleural friction rub. What is more, auscultation may reveal pleural effusion, a collection of fluid between the parietal and visceral layers of the pleura. Diagnosis is established thanks to symptoms and signs, clinical findings and laboratory tests. Imaging studies such as CT pulmonary angiography are of additional help. How is Pulmonary Embolism Treated? The cornerstone of treatment is anticoagulant therapy. Anticoagulants dissolve the blood clot and establish reperfusion of the affected lungs. Symptomatic and supportive treatments are applied too. Initially, practically all patients are administered heparin or fondaparinux. After a few days patients continue taking other anticoagulants such as warfarin, acenocoumarol or phenprocoumon. During the treatment all patients are closely monitored and parameters regarding coagulation are frequently checked. The dose of the drug is always adjusted according to laboratory findings. Treatment is administered in hospital environment while only when bodily functions are normalized patients may continue taking medications at home. Apart from treatment for pulmonary embolism it is essential to treated deep vein thrombosis as well and prevent recurrent obstruction of lung blood vessels. Finally, prognosis of pulmonary embolism practically depends on the portion of the lungs affected by obstruction. Comorbidities make the prognosis worse. Massive pulmonary embolism easily leads to death if anticoagulants are not administered straight away. Chronic embolisation is a common trigger of pulmonary hypertension. All in all, the condition can be treated and patients saved on time. The problem is that once anticoagulant therapy

is discontinued the recurrence of deep vein thrombosis and its complication - pulmonary embolism rises.

PULMONARY VASCULAR DISEASES. pdf

8: Pulmonary vascular disease - ERS

The pulmonary vascular disease program provides expert care for patients with this type of advanced lung disease. Diagnostic and management services are available for any of the following hypertension conditions.

What Is Pulmonary Hypertension? Pulmonary Hypertension PH is high blood pressure in the blood vessels of the lungs. It occurs when the small blood vessels that go through the lungs become thicker, constrict tighter or become plugged. This, in turn, leads to increased pressure in those vessels making the right side of the heart work harder to pump blood through the lungs. If high pressure remains, the heart will become enlarged and weaker, pumping blood less efficiently into the lungs and eventually throughout the body. Patients develop progressive fatigue and shortness of breath, two typical symptoms of PH. Given the complexity of some of the drugs and how they are given, it is very important that physicians who can diagnose and treat PH with the full range of treatment options evaluate patients at a pulmonary hypertension center. What are the symptoms of pulmonary hypertension? Early in the disease, symptoms may be nonspecific and can mimic symptoms of other medical conditions. It is also possible that patients may experience only limited symptoms. The following are some of the most common symptoms of PH: Worsening shortness of breath also called dyspnea Fatigue or tiredness Chest pain or pressure during activity angina Dizzy spells that may occur during activity or exercise Fainting Ankle or Leg swelling edema Increased heart rate tachycardia Cough What causes PH? While we do not yet know the causes of PH, we do know that it affects people of all ages, races and ethnic backgrounds. Likewise, patients with valvular or hypertensive heart disease, or severe lung disease can also develop different forms of PH. We are experienced in treating all forms of pulmonary hypertension. Numerous treatment options for pulmonary hypertension are available, depending on the type of pulmonary hypertension. Treatments for pulmonary arterial hypertension range from medications to transplant surgery. Our researchers are also studying new treatments for PAH all the time. Medications Several types of medications are used to treat PAH. Prostacyclin analogues facilitate the vessels in the lungs to open more and allow blood to move through them with less resistance vasodilation. These drugs may be given by continuous IV infusion, infusion under the skin, or as an inhaled therapy. We are exploring other delivery methods for prostanoids, including oral administration. Endothelin receptor antagonists, in pill form, help reverse the effect of endothelin, a substance in blood vessels that causes constriction. Phosphodiesterase-5 inhibitors relax the blood vessels in the lungs. Anticoagulants help prevent the blood from clotting. Diuretics help the body remove excess fluid that may accumulate in tissues due to high pressures in blood vessels. Antiproliferative agents are anti-cancer drugs that may have a role in treatment of PAH by reducing abnormal growth of the cells that line the lung arteries. Many drugs for treating pulmonary hypertension have side effects. The dosage level must be carefully set and monitored to avoid serious side effects or complications. We work with patients to find the most effective and safe doses. Chronic Thromboembolic Pulmonary Hypertension Chronic thromboembolic pulmonary hypertension is a rare form of both pulmonary hypertension and blood clots that go to the lung. Our pulmonary vascular team has experience in diagnosing and in both the medical and surgical treatment of this condition. Some, but not all, patients have a history of blood clots to the lung. Correct diagnosis is important because in many cases a surgical procedure called pulmonary thromboendarterectomy can successfully treat this condition. This procedure may be recommended for people whose secondary pulmonary hypertension is due to persistent pulmonary emboli blood clots. Follow-Up Care We not only diagnose and treat pulmonary hypertension, we also provide long-term care and support to help patients manage their condition. Our team of physicians and nurses develop individualized treatment plans and follow up with each patient on a regular schedule. Patients also can access information about research opportunities and continuing education. Close follow-up care helps physicians detect early changes that may require further treatment.

PULMONARY VASCULAR DISEASES. pdf

9: Lung Institute | Pulmonary Vascular Disease

Pulmonary vascular disease; Pulmonary vascular disease Supplementary Material. kB. WB PVD Supplement. The burden of lung disease. Introduction: the global.

Bronchiectasis Upper airway obstruction Patients with small airway obstruction, BO, and follicular bronchiolitis usually present with a non-productive cough, wheezing, and dyspnea. These symptoms may be exacerbated by triggers similar to those of a patient with asthma. Patients with BO may also present with more rapidly progressive symptoms than are typically seen in small airways disease. In RA, the disease tends to correlate with joint symptoms, but it has been described prior to the onset of articular manifestations. Patients complain of a productive cough and progressive dyspnea on exertion. These symptoms are exaggerated during an acute infection, at which point patients may present with increasing or changing sputum production and hemoptysis. Crackles are often heard on examination, although a small percentage of patients may present with wheezing due to airway hyperresponsiveness. RA and SLE patients who present with hoarseness, dysphagia, or pain with speaking or swallowing should be evaluated for an upper airway obstruction caused by cricoarytenoid arthritis. Knowing these early symptoms may prevent the patient from developing stridor, difficulty breathing, and a potentially life-threatening upper airway obstruction requiring emergency tracheostomy. Pleural disease Pleural effusions Table 3 and pleural inflammation or pleurisy are the most common pleural complications associated with CVD. In SLE, the effusions, which are frequently accompanied by pleuritic chest pain, are usually small and bilateral. Fever and tachycardia can accompany the effusion, raising the concern for infection. Thoracentesis findings in SLE are consistent with inflammation but not very specific. The appearance can range from serous to frankly hemorrhagic. They have low pleural fluid complement levels, and LE cells neutrophils or macrophages that have engulfed nuclear material of other cells may be seen. Again, these findings are not specific. They are usually small and unilateral, often occur later in the disease course of RA, and occur more often in men and in those with high rheumatoid factor RF titers. Up to half of the time, they occur in those with subcutaneous nodules. Findings on thoracentesis more suggestive of RA include high protein levels, significantly reduced glucose levels, elevated lactate dehydrogenase, and a moderately depressed pH. The RF levels of the pleural fluid are often equal to or even greater than those found in the serum. Pulmonary vascular disease The most common vascular complication is secondary pulmonary hypertension related to parenchymal disease, primary pulmonary arterial hypertension or pulmonary venoocclusive disease, especially in scleroderma. Patients with pulmonary hypertension may present with generalized fatigue and progressive dyspnea on exertion. They may also complain of exertional chest pain or syncope. Early on, the only physical examination finding may be an augmented second heart sound. Later, the patient may have a systolic ejection murmur or a diastolic regurgitation murmur. Patients may also demonstrate signs of heart failure, including lower extremity edema or abdominal tenderness from hepatic congestion. Vasculitis, including necrotizing pulmonary capillaritis, is a rare complication of CVD and seen most commonly in SLE. Vasculitis and capillaritis should be considered in patients who present with diffuse alveolar hemorrhage. Pulmonary embolism occurs with increased frequency in patients with SLE and antiphospholipid antibody syndrome. These patients are at risk for chronic thromboembolic pulmonary hypertension. Respiratory muscle weakness Patients with respiratory muscle weakness present with dyspnea on exertion and orthopnea. Orthopnea reflects further impairment in diaphragmatic function in the supine position with the loss of the assistance of gravity and the shifting of the abdominal contents cephalad. Associated involvement of the proximal striated muscle portion of the esophagus can cause oropharyngeal dysphagia and lead to a risk of aspiration pneumonia. Shrinking lung syndrome is an ill-defined respiratory muscle disorder associated with SLE. In addition to dyspnea and orthopnea, patients with this disorder often have episodes of pleuritic chest pain. Shrinking lung syndrome is believed to be due to diaphragmatic dysfunction, but whether this is the result of a primary myopathy of the diaphragm or phrenic neuropathy is unclear. Miscellaneous disorders Acute reversible hypoxemia syndrome is a rare complication of SLE. While the pathogenesis remains unknown, it is believed to involve upregulation of the complement cascade leading

to a leuko-occlusive vasculopathy. It is a diagnosis of exclusion in patients presenting with acute hypoxia without evidence of parenchymal abnormalities on chest imaging or venous thromboembolism. Treatment includes corticosteroids with or without aspirin. Patients with RA can develop pulmonary nodules that typically correlate with subcutaneous nodules identified in other locations. These nodules tend to be subpleural and subcentimeter in size. A patient may present with a solitary pulmonary nodule or multiple nodules that wax and wane, and central cavitation can occur. These nodules should be followed to ensure there is no underlying malignancy. Many rheumatoid nodules regress spontaneously or with treatment of the underlying CVD without significant symptoms or complications. In addition to pulmonary toxicity due to underlying CVD, many immunosuppressive and biologic agents used to treat the manifestations of CVD are associated with pulmonary toxicity ranging from infections to pneumonitis and should be considered in the differential of CVD patients with pulmonary symptoms. Secondary infections, often related to immunosuppression, can mimic a number of pulmonary complications of CVD and efforts should be made to identify these given the treatment implications. Rarely malignancies can also present similarly to pulmonary complications of CVD and if suspected, biopsies should be pursued. Lymphocytic interstitial pneumonia LIP can present with centrilobular or subpleural nodules. Although LIP represents a benign lymphoproliferative disorder, it is important to document polyclonality with immunohistochemistry to distinguish nodules related to LIP from lymphoma. Not applicable Which individuals are at greatest risk of developing a pulmonary complication of a collagen vascular disease? ILD has become the leading cause of death in patients with scleroderma, especially diffuse cutaneous scleroderma. The association between tobacco use and ILD is less clear than once reported. Airways disease The risk of developing small airways obstruction and BO is greatest for patients with RA and scleroderma. Esophageal dysmotility and gastroesophageal reflux disease can increase the risk of recurrent aspiration and subsequent bronchiectasis in scleroderma. Upper airway manifestations of RA, including cricoarytenoiditis and cervical arthritis, are more common in females. Patients with SLE and associated antiphospholipid antibody syndrome are at greatest risk for pulmonary embolism or diffuse alveolar hemorrhage from vasculitis. Shrinking lung syndrome is seen only in patients with SLE. What laboratory studies should you order to help make the diagnosis, and how should you interpret the results? Interstitial lung disease Some serologies support the presence of an underlying CVD and may also predict the presence of ILD as a complication. Anti-Jo-1 is anti-histidyl-tRNA synthetase that can be seen in the anti-synthetase syndrome. This syndrome consists of myositis, ILD, arthritis, and fever, although not all components of the syndrome are always present. The other anti-synthetase antibodies are associated with varying parts of the triad of the syndrome ILD, arthritis and fever and some occur in patients with ILD alone and thus a high index of suspicion is necessary to diagnose them. Patients with antibodies against melanoma differentiation-associated gene 5 anti-MDA-5 can present with rapidly progressive ILD and skin ulceration in the absence of other dermatologic or myopathy symptoms. Anti-Scl is a breakdown product of topoisomerase I. Specific for scleroderma, it is more common in diffuse scleroderma than limited disease. In various studies, having a positive Scl increased the risk of having ILD from one to five times over that of those without it. In addition, having a positive Scl is associated with increased mortality. Its counterpart is the anti-centromere antibody that is also specific for scleroderma but is more common in limited disease and is more often associated with pulmonary hypertension. RF is made up of antibodies against the Fc portion of IgG. These antibodies are fairly non-specific and are most often associated with RA. What imaging studies will be helpful in making or excluding the diagnosis of a pulmonary complication of a collagen vascular disease? Interstitial lung disease Plain CXRs may demonstrate alveolar, interstitial, or focal consolidative changes, depending on the underlying pattern of ILD. Honeycombing may be evident in advanced disease. The CXR is relatively insensitive and non-specific. Airways disease Patients with small airways disease do not usually require further chest imaging unless they do not respond to usual therapies. Patients with CVD may have had chest imaging for other reasons. A CXR can show hyperinflation and mild bronchial wall thickening in the presence of airways disease, while a chest CT may show bronchial wall thickening, air-trapping, and bronchiectasis. Patients with BO may have normal chest imaging so the diagnosis should be considered in a patient who has not responded to typical therapies for small airways disease and who has air-trapping or other signs of small

airways disease on HRCT. Because air-trapping can help narrow the differential of a patient with CVD who presents with a cough or dyspnea, inspiratory and expiratory images should be obtained. Bronchial wall thickening or bronchiectasis may also be seen. Chest CT images of patients with follicular bronchiolitis will show centrilobular and peribronchial nodules. Bronchiectasis, which should be evident on chest imaging, is defined as thickening and dilation of the airways. Airways are considered dilated if they are larger than their adjacent pulmonary artery. CT imaging of the neck may demonstrate cricoarytenoid abnormalities in patients with RA and upper airway obstruction. Pleural disease Pleural effusions in SLE are typically small and bilateral, but moderate to massive effusions have been described. In RA effusions are also typically small and unilateral, but they can vary widely in size. Very small effusions can easily be missed on plain CXR; chest CT is more sensitive for detecting small effusions. Ultrasound can be useful in diagnosing the presence of an effusion and also in preparing for a thoracentesis by identifying an appropriate site. Pulmonary vascular disease Pulmonary hypertension may be suspected if the main and central pulmonary arteries are enlarged on CXR or chest CT. Dilation of the right atrium and ventricle may also be apparent. CT imaging may reveal a mosaic attenuation pattern, with decreased attenuation in areas of vascular dropout. Expiratory images may be helpful in ruling out air-trapping. As CT findings are relatively insensitive for pulmonary vascular disease, a transthoracic echocardiogram should be ordered in patients when diffusing capacity of the lung for carbon monoxide DLCO is reduced out of proportion to spirometry, in those with lightheadedness or dizziness, and annually in those with scleroderma to assess for pulmonary hypertension. Patients who present with diffuse alveolar hemorrhage from capillaritis or vasculitis usually have bilateral airspace opacities on CXR. On CT scan, multifocal ground-glass opacities or consolidation are present. Respiratory muscle weakness The presence of small lung volumes on CXR and chest CT in the absence of ILD is a clue, albeit a nonspecific one, to the possible presence of respiratory muscle weakness. There may be associated basilar atelectasis that can sometimes be mistaken for ILD. Fluoroscopic inspection of the diaphragm during a rapid inspiratory maneuver "sniff test" can be helpful in documenting unilateral diaphragmatic weakness, but false negative studies are common when there is bilateral diaphragmatic weakness.

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