**Pathology Cardiovascular Paragraph**

![Ischemic Heart Disease Diagram]

**Atherosclerosis.** This is a process in which deposition of cholesterol and fat into the walls of a vessel cause a yellow, fatty lesion. Atherosclerosis commonly occurs on the larger vessels (aorta, carotid, coronary, popliteal) while arteriolosclerosis occurs on smaller vessels. Risk for disease is increased with smoking, hypertension, diabetes, ↑Cholesterol, and ↑Fat (aka a typical American). The lesion consists of a fatty atherosclerotic plaque on the lumen wall. This is the primary lesion, developed from a fatty streak. Everybody has these fatty streaks because everyone has some level of cholesterol and fat in their blood. But, what happens is that smooth muscle cells migrate around it, act as fibroblasts and lay down a fibrous plaque. This fibrous cap protects the lesion, keeps it away from growing and injury. This usually causes no problems unless it consumes 95% of the lumen. What happens in atherosclerosis is that the lesion ruptures, exposing collagen, vWF, or the cholesterol to platelets. This is called a complex atheroma. This is where a thrombus gets built on top of the plaque, causing occlusion of the vessel, or the cholesterol can break off and effect distal structures. Atherosclerosis predisposes the patient to Abdominal Aortic Aneurysms, Ischemic Events (heart, but all organs can be effected), Peripheral Vascular Disease (pain in calf when patient walks), thrombus and emboli.

**Arteriolosclerosis** is the condition of the smaller arteries under various stressors. For example, hyperplastic arteriolosclerosis is the onion skinning of malignant hypertension while hyaline arteriosclerosis is the pink, glassy material associated with diabetes or benign hypertension.

**Essential Hypertension.** A whole lot of people have hypertension (about 25% of the US population) with the largest incidence in the African American population. Of everyone who has hypertension, only 10% are known WHY. Thus, Essential HTN, or Primary HTN, is in fact idiopathic. This is an asymptomatic disease (the patient doesn’t know they have it until you take their blood pressure). Long term, hypertension causes a heap-load of problems. Look for hyaline arteriosclerosis on small vessels. Hypertension increases the risk of atherosclerosis, dissecting aneurysm, and coronary heart disease, and ventricular hypertrophy/failure. Every organ is affected by hypertension.

**Secondary Hypertension.** This is far less common than essential hypertension. It is “renal hypertension.” For whatever reason it may be, the kidneys sense a decreased flow through the glomerulus (this may be because of renal artery stenosis or because the kidneys are just broken). The kidney responds by activating the renin-angiotensin system. Renin turns on ANG-1, ACE turns on ANG-II. ANG-II has two effects. Its first is to activate aldosterone who conserves Sodium, and with it water, expanding the extracellular fluid volume (fluid overload). The second effect is to cause direct vasoconstriction of vessels leading to an increased peripheral vascular resistance.
**Malignant hypertension.** An acute condition whereby the blood pressure is either greater than 200 systolic or greater than 120 diastolic. The patient will experience blurry vision, and is at increased risk for cerebral hemorrhage. Blood vessels (particularly the vulnerable kidney) will demonstrate onion-skinning called hyperplastic change. The kidneys will have a flea-bitten appearance on the surface of the cortex from all the petechial hemorrhages. The eyes exhibit flame-tip lesions or papilledema. This is a medical emergency and must be corrected immediately. You will learn more of this when we discuss the Pharmacology of depression (combining MAO-Is and TCAs can cause an undesired effect).

**Stable Angina.** The patient has so much atherosclerosis that 75% of the blood flow is occluded to the coronary artery. When they exercise, cardiac demand increases, but the lumen is so narrowed, not enough blood can be pumped to the myocardium. This causes a transient Subendocardial ischemia producing an ST segment depression (the epicardium gets a lot of blood and steals the oxygen going to the distal endocardium) that is relieved with rest or nitrates. This will progress to unstable angina unless the patient does something about their diet, exercise, and smoking.

**Unstable Angina.** The most important Angina is unstable angina. It is also called a crescendo angina, “crescending” to the MI. This is a severe atherosclerotic plaque (90% occlusion) that then ruptures causing a superimposed thrombus on top. This causes chest pain at rest that is not relieved by nitrates or rest. There is very little differentiating this picture from a full myocardial infarction. In fact, you can look at this as the start of an MI, just before the heart muscle actually dies. It is the ischemia before the necrosis. You pick this over an MI when the patient complains of increased frequency or duration of chest pain without rest that subsides spontaneously. In practice, there is no way of knowing if THIS chest pain RIGHT NOW is unstable angina or a full MI. There will be ST Segment Elevation (transmural).

**Prinzmetal Angina.** This is a chest pain that is not associated with exercise, stress, or atherosclerosis. Prinzmetal Angina is a coronary vasospasm that transiently obstructs flow. It can be just as damaging and deadly as any other occlusion, but is more problematic because it is not relieved with rest and nitrates. The only thing we can do is attempt to vasodilate the coronary artery (which is difficult to do because there is no sympathetic innervation to the coronary arteries). It looks like an MI because there is ST segment elevation.

**Myocardial Infarctions.** An MI is defined as a local area of necrosis of cardiac tissue due to ischemia. It is the number 1 killer in the United States. There are two subtypes of myocardial infarction. Because the coronary arteries traverse the external side of the heart and send capillaries into the myocardium from the outside, the “inside” (i.e. the endocardium) is farthest from the blood supply. In a state where there is minor obstruction to flow, the “outside,” that is, the epicardium, will be supplied while the endocardium is not. A Subendocardial infarction does not transverse the entire wall and is demonstrated by ST Segment Depression on the ECG, usually a result of moderate decrease in flow. A Transmural Infarction does transverse the entire wall of the heart, is demonstrated by a ST Segment Elevation on the ECG, and is the result of severe decrease in flow. Because of the setup of the arteries of the heart, you can assume that there are no collaterals in the heart (in reality there are, but for Test taking simplify your world by saying there are none). That means if one artery becomes occluded, everything distal to it will become ischemic. The most common ischemic event is occlusion of the **Left**
Anterior Descending Artery, which supplies the left ventricle. An infarction of the left ventricle causes an immediate decrease in cardiac output, which can result in cardiogenic shock, death, and hypoperfusion of other end organs. During an ischemic event, the gross and histologic changes are important to recognize, as are the dangers of an infarction. In the first day, the tissue dies. There is no gross change but there is evidence of Coagulative necrosis (myocytes without nuclei) with the earliest sign being wavy degeneration in 1-4hrs. After the first day, neutrophils will enter (myocytes without nuclei surrounded by multi-lobed leukocytes) to eat away the dead tissue. Grossly, there is a hyperemia around the infarct site. From the start of the MI to this point, there is a high risk of arrhythmia (oxygen has been deprived, ATP isn’t made, Na/K-ATPase cannot run, and electrolyte disturbances can cause arrhythmias of any kind). Arrhythmia is the primary cause of death in the first week. As neutrophils and macrophages begin to degrade the necrosis, the space is replaced by granulation tissue via fibroblasts. As the granulation tissue is laid down, it is at its weakest. Because the heart still beats despite its injury, the heart can literally tear itself open. After the first week there is a large risk of free wall or papillary muscle rupture. Both of these can be fatal (cardiac tamponade from free wall rupture and mitral insufficiency from papillary rupture), as a reduced cardiac output caused global ischemia. The granulation tissue works its way from the outside in, looking yellow in color. Finally, after many weeks to months, the granulation tissue is laid down and is firm. The contracted scar looks grey-white on gross. The rest of the muscle relaxes and contracts, but the scar tissue does nothing. This predisposes for the development of ventricular aneurysm and subsequent mural thrombi. The aneurysm is not like a berry aneurysm that ruptures (like in the brain), but rather an outpouring with paradoxical motion on an echo, a region of stagnant blood that reduces cardiac output and increases the risk of clotting. An arterial clot from a mural thrombus can cause infarction distally in the body (in the brain and kidney would be most fatal). Myocardial infarctions are suspected based on clinical picture (chest pain radiating to the jaw and down the right arm), screened for by ECG (ST segment depression is Subendocardial, ST Segment elevation is transmural, Q wave is an old infarct), and confirmed by enzyme levels (Troponin I is the best, CK-MB is next best, AST and LDH are unreliable). Sequella of a myocardial infarction are abundant, but watch out for a fibrinous pericarditis (Dressler Syndrome) and Heart Failure.

Left Ventricular Failure. Left ventricular failure is the result of either long standing hypertension or an acute myocardial infarction. In the first case, long standing hypertension induced an increased afterload that the left ventricle was forced to overcome. Whatever the cause of the hypertension is irrelevant (coarctation, essential, secondary), just that the heart had a larger pressure to push against. Well, the reaction of the heart to an increased workload is to hypertrophy. Sustained hypertrophy causes a wearing of the PKA intracellular signaling system, inducing a plastic change in the myocytes. Essentially, the heart gets bigger (and stronger), bigger, bigger, and then craps out. As its getting bigger you are likely to hear a 4th heart sound. After it craps out, you are likely to hear a 3rd heart sound. A left ventricular failure means that it cannot pump blood like it used to. Blood cannot get out of the heart and into the aorta, resulting in a decreased perfusion of end organs in the periphery, but also a build-up of fluid in the lungs called pulmonary edema. Hemosiderin-Laden Macrophages (alveolar macrophages with decomposed red blood cells) are pathognomonic for pulmonary edema. This back up of blood in the lungs increases the afterload for the right heart, leading to a right ventricular failure. While a heart
can fail chronically after prolonged hypertension and hypertrophy, an acute myocardial infarction can produce the exact same phenomena – decreased cardiac output. On a final note look for “boxcar nuclei” (cells get big and look like railroad cars) on histology, a product of hypertrophy.

**Right Ventricular Failure.** Right ventricular failure is very much like left heart failure. Either the afterload is too great or the there is an infarction of the right ventricle. The effects, however, are very different. If there is a right heart failure (only) the **venous return** to the heart cannot be tolerated. The right ventricle will not pump enough blood into the lungs. If not enough blood gets into the lungs, not enough blood gets into the left heart. If not enough blood gets into the left heart, not enough blood can get out in the periphery (ah, good ol physio). Because the veins are big and floppy, all the blood gets stored in the veins. This means there is **no pulmonary edema** as there was for LHF, but there will be significant **venous congestion**. This is identified by an **elevated jugular veins point** (you can see the vein bulging from the neck) and from **hepatomegaly**. Since the blood is building up, and gravity takes it down, the fluid accumulates really in two places – the liver and the legs. Thus you get **peripheral edema** and a **nutmeg liver**. The increased pressure in the liver causes a decrease of blood flow, resulting in **Centrilobular necrosis** of the liver giving it the appearance of a nutmeg on cross section. While Left Heart Failure and Right Heart Failure commonly occur together, any test will want you to be able to separate them. So, even though the **most common cause of RVF is LVF**, look for reasons like **Cor Pulmonale** (primary pulmonary hypertension), pulmonary artery stenosis, or a congenital heart defect.

Really what you want to know is that **Left heart** failure causes a decreased cardiac output and pulmonary edema, the result of an infarction or long term systemic hypertension while **Right heart** failure causes a decreased cardiac output, peripheral edema, nutmeg liver, and is the result of pulmonary hypertension.

**Left To Right Shunts.** Left to right shunts are characterized by the left ventricle pumping oxygenated blood into both the systemic and the pulmonic vasculature, leading to a non-cyanotic deficiency. Babies are pink, though their perfusion may not be great. These all result in a pulmonary hypertension and right ventricle hypertrophy with the eventual reversal of the shunt leading to cyanosis late in life (called **Eisenminger’s syndrome**). All Left → Right Shunts have a “D” in their name.

**Atrial Septal Defect** (ASD). This is the most common congenital defect seen in adults only. It commonly occurs on its own (90%) and, if with another abnormality, it is usually that abnormality that is more hemodynamically significant. Essentially, there is a hole between the left and right atrium. There are three kinds: **Secundum** (90% of cases), **Primum** (5% of cases), and **Sinus Venosus** (5% of cases). The
blood is shunted to the right because the right ventricle has greater compliance and can accept the excess fluid. Since the pulmonary vasculature receives more fluid, it is under greater tensions, and therefore there is pulmonary hypertension. From the excess fluid and pulmonary hypertension, the right ventricle hypertrophies. There is a systolic murmur associated with the shunt. In the end, the right ventricle gets strong enough that the shunt will reverse leading to cyanosis (R→L shunt bypasses lungs).

**Ventricular Septal Defect** (VSD). This is the most common congenital cardiac defect overall. It rarely occurs on its own (30%) and is often associated with another syndrome (such as Tetralogy). There is a hole between the ventricles, and come in two types: Membranous (90% of cases) and Infindibular (10% of cases). Blood is shunted from left to right because of the more forceful left ventricular contraction sending blood directly into the right ventricle and pulmonary vasculature. This again leads to pulmonary hypertension and right ventricular hypertrophy. Pulmonary Hypertension eventually results in Vasculitis, significantly increasing the resistance, where the shunt reverses leading to cyanosis. Most VSD close on their own, so surgery is not attempted until approximately 1 year of age.

**Patent Ductus Arteriosus** (PDA). The Ductus Arteriosus is the shunt from the pulmonary artery to the aorta distal to the major branches (carotid and subclavian). Embryologically, it bypassed the fetal lungs, but should close shortly after birth. Closure is mediated by an absence of prostaglandin E and by increased oxygen tension. A patent ductus in the born infant allows for left to right shunt, whereby the powerful left ventricle shoots blood into the systemic and pulmonary vasculature directly. This shunt can eventually reverse as well. Use of the NSAID indomethacin prevents prostaglandin synthesis and results in closure (from First Aid, “ENDomethacin ENDS the Ductus”). Misoprostol (PGE) maintains the patency of the ductus, as may be required in certain other abnormalities (such as infantile coarctation). The buzzword for PDA is a continuous and/or machine like murmur.

**Right to Left shunts.** So-called blue baby syndromes are where the right ventricle is allowed to pump deoxygenated blood into the periphery, leading to cyanosis. These shunts do not reverse, and are often fatal unless corrected surgically (Cyanosis doesn’t do so well for, say, brain development). All these disorders have a “T” in them.

**Tetralogy of Fallot.** You gotta know this one. It is characterized by four things: (1) VSD (2) Subpulmonic Stenosis with right ventricular outflow obstruction (3) An aorta that sits on top of the VSD, and (4) right ventricular hypertrophy. The degree of symptoms is dependent on the degree of stenosis. If mildly stenosed, you may have a pink Tetralogy where there is actually a left to right shunt, as the outflow of the right ventricle is not totally obstructed. However, as the infant develops, the aorta gets larger and pushes the pulmonary artery out of the way, so that all babies will become blue babies. This is caused by the deoxygenated blood being obstructed from entering pulmonary vasculature, and is instead shunted into the left ventricle and into the periphery. This is all caused by an anterosuperior displacement of the infundibulum. Note that if there were complete right ventricular obstruction, there would have to be a patent ductus to allow any oxygenation of the blood (as in pulmonary stenosis/atresia), suggesting that some blood is oxygenated. The patient may present with cyanosis that resolves with squatting, prompting an X-ray or CT revealing a boot shaped heart.
Transposition of Great Arteries. This is literally a swapping of the pulmonary artery and aorta. The left ventricle is connected to the pulmonary artery and the right ventricle is connected to the aorta. This is caused by a failure to spiral during development. This effectively creates two circulations: (1) Left Atrium, Left Ventricle, Lungs and (2) Right Atrium, Right Ventricle, Systemic. The left side pumps and receives oxygenated blood to and from the lungs. The right side pumps and receives deoxygenated blood to and from the periphery. Without any other defect, this disease is incompatible with intrauterine life. There must be some connection between left and right systems (PDA, VSD, ASD) that allows mixing of blood. This requires surgery to fix. Obviously there will be a right ventricular hypertrophy and a hypoplastic left ventricle. Associated with Diabetic Mothers (that is poorly controlled diabetics who get pregnant, not women who develop diabetes during their pregnancy. The development of the heart occurs during week 4).

Truncus Arteriosus. This is a failure of the common tube to separate into the pulmonary artery and aorta. That means one giant trunk receives blood from both the right ventricle and left ventricle, going to both the systemic and pulmonic vasculature. This is half normal: The right ventricle pumps to the lungs, while the left ventricle pumps to the system. This is half abnormal: The right ventricle pumps to the systemic vasculature (causing right ventricular hypertrophy and cyanosis) while the left ventricle pumps to the pulmonary vasculature (pulmonary hypertension exacerbating RVH).

Obstructive Diseases do not cause cyanosis or pulmonary hypertension. Their effects are dependent on where they are, but they all cause an obstruction to flow of blood in general, which is bad.

Coarctation of the Aorta. Most important of the obstructive diseases. By definition, this is a narrowing of the aorta from birth not associated with atherosclerosis, but a congenital anomaly. There are two types. Infantile type has a coarctation proximal to major branches, limiting blood flow to, well, everything, relying on a patent ductus for survival. This you see right away and requires immediate intervention. The patent ductus delivers less oxygenated blood to the system, including the brain. Adult type can present at any time in life, and is a coarctation distal to major branches. This presents with hypertension in the upper extremities (increasing risk for dissection, cerebral aneurysm, and embolism) with hypotension in the lower extremities (weak pulses, claudication with exercise). Collateral circulation through the thoracic arteries causes engorgement which subsequently erodes the ribs resulting in rib-notching on X-ray. Surgery has very successful outcomes. However, if caught too late, perceived increased vascular resistance may have already induced hypertrophy or failure. There is a high association with Turner Syndrome (45,XO).

Pulmonary Atresia. This is an isolated stenosis or failure of the pulmonic valve to recanalize. This limits right ventricular outflow. With complete obstruction there is a hypoplastic right ventricle and the requirement for some septal defect for life. If significantly atretic, right ventricular hypertrophy results and Vasculitis from the jet stream through the atretic valve results in pulmonary hypertension. This bares a poor prognosis.

Aortic Atresia. This is an isolated stenosis or failure to canalize of the aortic valve. Complete atresia results in hypoplastic left heart syndrome and requires a PDA for survival. Forms of atresia include aortic stenosis and bicuspid aortic valve.
**Infectious Endocarditis.** This is a disease caused by bacterial (or fungal) vegetations on the myocardium and valve leaflets. There are two types. **Acute Endocarditis** is marked by a high fever (chills, nausea, fever), large vegetations that can ulcerate to form ring abscesses with a rapid onset, usually associated with *staph aureus*. Even with treatment, acute endocarditis is 50% fatal. **Subacute Endocarditis** may hide behind a low grade fever, is not nearly as virulent (no abscesses), has a slower onset, and is usually associated with *strep viridians* (dental organisms). Regardless of the organism, the vegetations occur on one side of the valve, may be singular or numerous, and all line up on the valve. These vegetations are friable and can break off to form septic emboli, causing septic infarctions in the brain, kidneys, and other organs (a major complication). Vegetations normally occur on the left side of the heart except in the cases of IV drug users where vegetations are introduced through venipuncture and are revealed on the right side of the heart. Endocarditis can occur on normal valves, but any modification that causes any abnormal flow (prolapse, stenosis, commissural fusion, atresia, bicuspid aortic valve, or even a prosthetic valve) increases the risk. Clinical identification is based on serologic and other lab tests, but the boards still test on the old indications (Osler Nodes, Janeway Lesions, and Roth Spots). Treatment centers around identification of the organism and susceptibility to drugs.

**Noninfectious Endocarditis.** In this disease, sterile vegetations line up along the valve leaflet. They can still cause emboli and infarction, but are sterile (full of fibrin) and therefore do not cause septic infarcts. It is attributable to a hypercoagulable state (cancer) and is often associated with bed-ridden elderly patients. It can be fatal, but the condition that predisposes them to the disease likely signifies a poor prognosis anyway.

**Libman-Sacks endocarditis.** This is the endocarditis of Lupus (“SLE causes LSE”). In particular, it is associated with the antiphospholipid portion of Lupus. These form sterile vegetations on both sides of the valve, involving the AV valves more than semilunars. There is a fibrinoid necrosis encircling the lesions and there may be hematoxilin bodies present.

**Rheumatic Heart Disease.** Stems from Acute Rheumatic Fever (RF). RF is a sequella of untreated Strep Pyogenes pharyngitis. It requires the infection, the lack of treatment, and a genetic predisposition for the disease. Antibodies to the M protein or pneumolysin O protein on the Strep cross react with heart tissue. Thusly, **anti-pneumolysin-O titer is elevated** (↑ASO). Rheumatic Heart Disease is diagnosed based on 5 major criteria (chorea, migratory arthralgia, Erythematous lesions, Carditis, and subcutaneous nodules) and some minor criteria (fever) that will resolve spontaneously. During the attacks, you must
identify the histologic specifics: Aschoff Bodies (which are diffuse patchy granulomatous infiltrates) and the present of Anitschkow Cells (macrophages). What is left over after recurrent attacks throughout a lifetime is a stenosed mitral valve. The valves are likely to fuse (fish-mouth or button-hole appearance). This affects the aortic and mitral valves, leading to left sided hypertrophy, failure, and subsequent CHF. Occasionally the myocardium may dilate, but most often it is a hypertrophy that leads to failure.

**Degenerative Aortic Calcific Stenosis.** This is an age-related aortic stenosis whereby calcifications adhere to the aortic valve, preventing its opening fully. This is no commissural fusion as with rheumatic disease. This affects the elderly in their 50s-70s and mandates an immediate surgery. Clinically, it is detected as a crescendo-decrescendo systolic murmur (as with all aortic stenosis).Concentric left ventricular hypertrophy is a compensatory mechanism to overcome the stenosis (increased resistance) which can progress to failure and CHF. If the stenosis is severe enough, the patient is at risk for syncope and sudden cardiac death.

**Mitral Annular Calcification (Stenosis).** Calcifications form in the annular ring of the mitral valve, usually slowly over decades as in Aortic Calcific Stenosis. These depositions may have multi-functional consequences dependent on where they are. They can cause insufficiency by preventing complete closure (mitral regurgitation) or they can cause stenosis by narrowing the valve. Mitral stenosis is a diastolic murmur caused by a restricted flow. Nodules of calcifications may impinge on the nodal conduction system leading to arrhythmias. Rarely, it leads to infectious endocarditis

**Mitral Valve Myxomatous Degeneration** (Mitral Valve Prolapse). This is the disease associated most often with mitral regurgitation (a harsh holosystolic murmur heard best at the apex of the heart). This is visualized by a ballooning mitral valve that penetrates back into the atria during systole. The Chordae can be stretched thin or can be fully ruptured. Microscopically, there is a thinning of fibrosa and a widening of spongiosa layers (fibrosa yields strength). This is common in the general population, and for most, causes no symptoms. However, there is an increased risk for thromboembolus and infectious Myocarditis requiring prophylactic antibiotics prior to dental surgery.

**Pericardial Effusion.** The pericardial sac contains between 20-50mL of fluid to decrease friction between the parietal and visceral layers. Some diseases can cause extra fluid to get into the pericardial sac. How fast it enters determines how well tolerated the additions will be. For example, serous pericardial effusions (commonly associated with CHF or hypoproteinemia) produce a small amount of fluid but can permit large volumes. Hemopericardium is a ton of blood in the pericardium without an inflammation reaction usually from a ruptured ventricular wall that is rapidly fatal. In between these two are a sero-sanguinous fluid caused typically by blunt force trauma. The real thing here is that you get constriction (really compression) of fluid the heart so it cannot relax, and you die from low ventricular output.

**Pericarditis.** This is an inflammatory reaction of the pericardial sac or either of its layers. It is almost always secondary to another disease nearby, and remits when the underlying cause remits. It has two phases: acute and chronic. Acute pericarditis is the active inflammation while the chronic pericarditis is the effect of the healing process. Which type you have will determine what the presentation is like and what the resultant effects will be.
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**Fibrinous Pericarditis** is the most important. It is associated with myocardial infarction, has a friction rub (muffled heart sounds), and is the most common. It is mostly fibrin leaking out after a transmural infarct that leads to irritation of the pericardium. When it heals it can lead to fibrinous strands that cause adhesive pericarditis.

**Purulent Pericarditis** is the worst one to have. This is caused by an infection that spreads to the pericardium by any route (direct contact, hematogenous, lymph). The infection itself can be cleared but the sequella are particularly nasty (adhesive and constrictive pericarditis).

**Hemorrhagic Pericarditis** usually follows cardiac surgery. This is like hemopericardial effusion except there is an inflammation reaction. This is different from a knife penetrating the myocardial wall or a rupture following an MI.

**Adhesive Mediastinopericarditis.** A healed form of purulent or even fibrinous pericarditis. The pericardial sac is obliterated and the heart forms adhesions to the mediastinum and associated visceral organs. Every time the heart contracts it has no smooth pericardial layer to reduce friction. Instead, it pulls against the lungs, the chest wall, the diaphragm, all just to beat. This requires severe hypertrophy, which will lead ultimately to failure.

**Constrictive Pericarditis.** A healed form of purulent pericarditis, the pericardium is stiff and calcified. It prevents diastolic relaxation of the heart and presents very much like restrictive cardiomyopathy. The pericardial sac must be removed in order to regain proper function.

**Dilated Cardiomyopathy.** Myopathies are problems with the myocardium itself, not secondary to any other disease process. In dilated cardiomyopathy there have been links to several causes: alcohol = thiamine deficiency and wet beri-beri; pregnancy; post-viral Myocarditis; Chagas Disease; and recently there has been some evidence of a genetic component. While there can be a mutation in any gene, the genes of the cytoskeleton seem to have the most impact the formation of dilated cardiomyopathy. The end result is a mild ventricular hypertrophy with significant dilation of all 4 chambers without any real usable histophysiologic changes. The dilation may involve a physiologic dysfunction of valves (dilation pulls the normal valves apart) but there will be a significant systolic failure with decreased cardiac output. This limits a healthy, young person from even running up or down the stairs.

**Hypertrophic Cardiomyopathy.** This is the disease that kills athletes on the court. There is a genetic defect in sarcomeric proteins (such as Beta-MHC) that alters contractile forces in the heart. Hypertrophic Cardiomyopathy is not a response to increased vascular resistance (as in ventricular hypertrophy following HTN), yet the heart is massive with hypertrophy. The heart muscle is so thick it cannot relax, and results in a diastolic dysfunction (the heart cannot fill). In gross, you see a bananalumen as there is an asymmetrical hypertrophy of the septal wall. The hypertrophy may be so great there is an additional systolic failure from an obstructed outflow. Histologically, the myocytes are in disarray, and are massively hypertrophic. This presents in an autosomal dominant inheritance pattern.
Restrictive Cardiomyopathy. In this cardiomyopathy “stuff” gets in the way of relaxation (diastolic failure). That “stuff” is usually amyloid, but can also be sarcoid or fibrosis. There are also subtypes of restrictive cardiomyopathy. Endomyocardial Fibrosis affects African boys and has little known about it except that fibrosis starts at the apex and rises to the mitral valve, often causing obstruction to outflow. Loeffler’s Endocarditis is associated with abnormal eosinophilia and potentially toxic degranulation, with a similar pattern of fibrosis as in endomyocardial fibrosis. This can be fixed by endocardial stripping, with total removal of the fibrotic layer. Fibroelastosis is a disease where there is a fibro-fatty coating to the entire endocardium that appears usually in children under 2 years of age.

Myocarditis. This is inflammation of the myocardium itself. It is most often associated with Cocksackie B virus, but other etiologies as well (such as Chagas Disease’s T Cruzi, Lyme Diseases B. Burgdorfi, or autoimmune reactions). It causes a 4-chamber dilation and may result in dilated cardiomyopathy. This predisposes for mural thrombi and a systolic failure. Histologic presentations depend on the organism doing the infecting. For example, Trypanosomes can be seen within myocytes in a T Cruzi infection. In viral infections there are patchy, diffuse fibrinous necrosis with inflammatory cell infiltrates (because they are diffuse, they are often missed by biopsy). In hypersensitivity reactions there is a perivascular infiltrate of lymphocytes and eosinophils, possibly leading to Vasculitis and myofiber necrosis.

Abdominal Aortic Aneurysm. This is a true aneurysm (involving all 3 layers of the artery) that almost always has atherosclerosis as a precursor. The abdominal aorta below the renal arteries has no vaso Vasorum, so relies on diffusion to irrigate the walls of the aorta. With a thick atherosclerotic plaque, blood does not make it, and the wall weakens. This is palpable as an abdominal mass and may be mistaken as a tumor. It requires surgery to fix as a large aneurysm may rupture leading to sudden cardiac death, as the entire cardiac output empties into the peritoneum. As it bulges it can theoretically collapse nearby vasculature (renal arteries), invade other tissue (intestines) and is a great spot for mural thrombi.
Syphilitic Aneurysm. This is caused by stage 3 syphilis. It has a preference for small arteries (such as the vasa vasorum). It causes an inflammation of the vasa vasorum, which results in occlusion and ischemic necrosis of the aorta. The muscle fibers are replaced with a fibrotic scar which is weaker than the muscle it replaced. This permits the aneurysm which often occurs in the ascending aorta. The bulging represent a threat of rupture, but most often presents with the signs of compression of nearby organs (dysphagia from the esophagus, dyspnea from the trachea of lungs, pain from vertebral erosion). You don’t really see this anymore since we treat syphilis or we give antibiotics for something else, which kills the syphilis. Involvement of the aortic root can cause aortic insufficiency with the diastolic murmur.

Aortic Dissection. This is where there is an intimal tear that penetrates but does not exceed the muscularis. It breaks the internal elastic lamina, but not the external. This forms a second cavity for the hematoma to build from. This presents as a sharp chest pain radiating to the back, and may descend as the tear descends. Instead of rupturing out into the peritoneum or mediastinum, it is possible that the vessel ruptures back into the lumen of the aorta (another intimal tear) forming a double-barrel aorta with two lumens as the tear endothelializes. Hypertension is almost always the precipitating factor (Marfan’s is next in line, if not hypertension). There are no warning sides of dissection other than the elastin fragmentation seen on histologic biopsy (which you wouldn’t do because you wouldn’t expect it). If the dissection returns to the aortic root there can be a problem with insufficiency.

Giant Cell (Temporal) Arteritis. This is the most common of the vasculitidies. It is characterized by head or facial pain from occlusion of the temporal arteries. A late complication is blindness (a medical emergency). The lesions are granulomatous with giant cells. The inflammation causes occlusion and ischemic damage. This disease responds well to steroids. Usually affects elderly females.

Kawasaki’s Disease. This is a disease that affects infants and neonates. It presents with a lymphadenopathy and/or a desquaminating skin rash caused by overproliferation of T-Cells. It can affect the carotid, leading to a possibly fatal complication. Usually self-limiting, rupture can lead to a fatal loss of blood.

Takayasu Disease. This is called pulseless disease and is endemic to Japanese Women. It causes stenosis of the aorta proximal to the major branches causing weak pulses in the extremities and decreased blood flow to the brain and kidneys. It is a disease that occurs towards middle age (30-40). Patients will present with neurologic or visual symptoms and will not have a radial pulse (or even a palpable carotid). Treat with steroids. It is rapidly fatal without intervention.

Wegner’s Granulomatosis. Granulomatic disease that affects the lungs, kidneys, and nose/sinus. It must be all three, and with all three is pathognomonmic for your purposes. It causes necrosis from ischemia though the etiology is not well understood. It affects medium to small arteries. It responds well to immunosuppression (cyclophosphamide). The C-ANCA is usually elevated (90% of patients). Do not confuse it for Goodpastures, which is hemoptysis and hematuria without nasal/sinus involvement (vignettes often sound similar). C-ANCA, Cyclophosphamide/Corticosteroids for C-Wegners (it’s so retarded you might just remember it that way).
Polyarteritis Nodosa (PAN). A poorly understood disease with granulomatous infiltrates forming randomly throughout the body. The symptoms are dependent on the organ affected. If multiple organ systems are affected in an “arterial way” than PAN should be expected. It causes a vascular necrosis and is often associated with Hep B. While not pathognomonic, also look for an elevated p-ANCA for PAN. Like most of the Vasculitis diseases, it responds well to steroids.

Buerger’s Syndrome. A disease that occurs in younger (30-40 year old), mediterranean chain smokers, in their extremities. There is a segmental thrombotic granulomatous disease that results first in Raynaud’s Symptoms followed by total occlusion and the pain of ischemic necrosis of the distal extremity. If it lasts long enough recanalization of the artery is possible. They must stop smoking.

Varicose Veins. A cosmetic problem of the superficial veins, usually of the legs. Commonly a result of valve failure, fluid accumulates in these superficial, noncommunicating veins that poses no threat to systemic vasculature. However, they are ugly, so people worry about them. Untreated, they can ulcerate causing great pain on the legs and limit movement. Risks for developing varicose veins are pregnancy, obesity, and a job where the legs are dependent (barber, surgeon).

Thrombophlebitis (deep vein thrombosis). Thrombosis we touched on last block. DVTs are increased by smoking, the pill, estrogen states, and coagulation disorders (Factor V Leiden). Because they occur in the deep veins, the potential for embolus (to the lungs) is high. Presents with pain in the calf with striking redness. Classic case is an elderly woman in a nursing home or a 16 hour flight where the patient doesn’t get up to move around. The patient presents either with difficulty breathing (pulmonary embolus) or pain in their legs (DVT that has not spread).