

Understanding Vascular Complications: A Primer of Essential Definitions

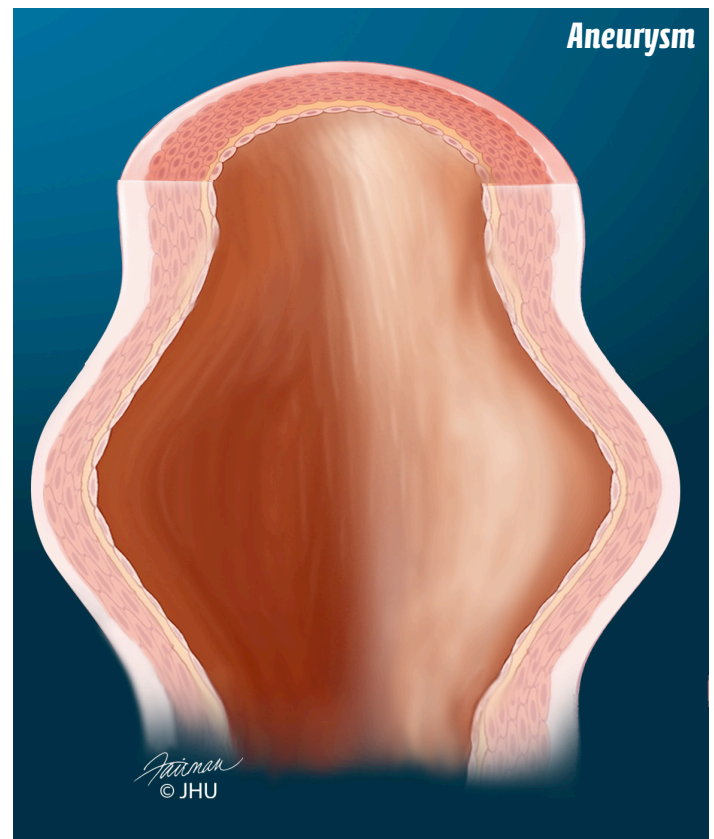
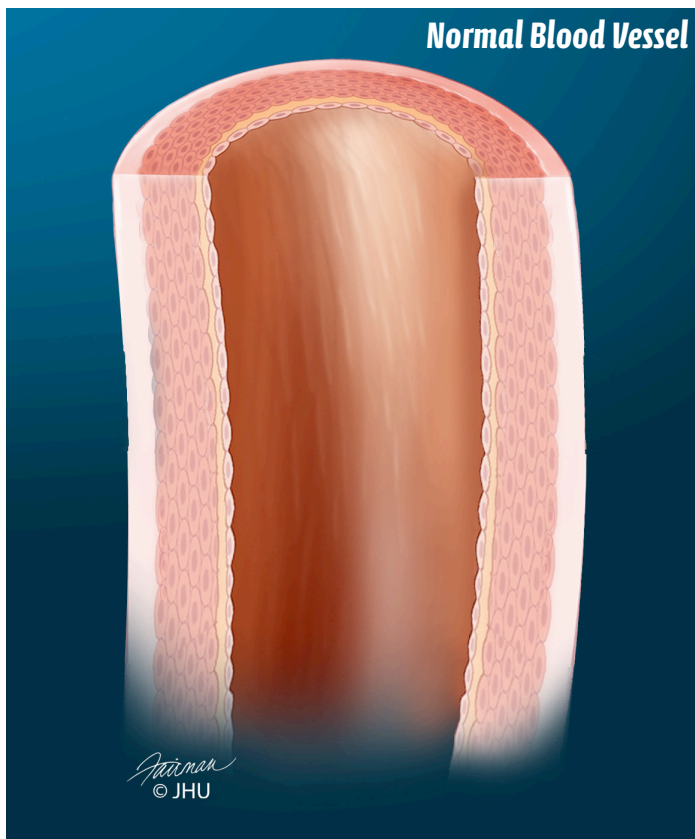
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ANEURYSM

All blood vessels consist of three layers, referred to as the intima, media, and adventitia. The intima is the innermost layer, and only a few cell layers thick. The media forms the middle layer and is mostly comprised of special muscle cells that provide elasticity. The outermost layer, the adventitia, is primarily connective tissue.

Blood vessels derive their strength from a web of structural proteins. The vascular complications of EDS arise from a genetic protein defect that predisposes blood vessel walls to be weak. True aneurysms occur in arteries and are defined as a dilatation of the blood vessel wall, but with all three layers intact. There is a special physics law which states that more stress is exerted on larger diameter blood vessels. This explains why as the diameter of arterial



aneurysms increases, they are more prone to rupture, in general. However, in EDS aneurysm rupture is unpredictable and may occur at any diameter. Aneurysm rupture is a life threatening condition, but fortunately true aneurysm formation is a relatively rare phenomenon in EDS, occurring in approximately 15% of patients. Aneurysms may be asymptomatic, or they may cause pain in the region of the aneurysm.

DISSECTION

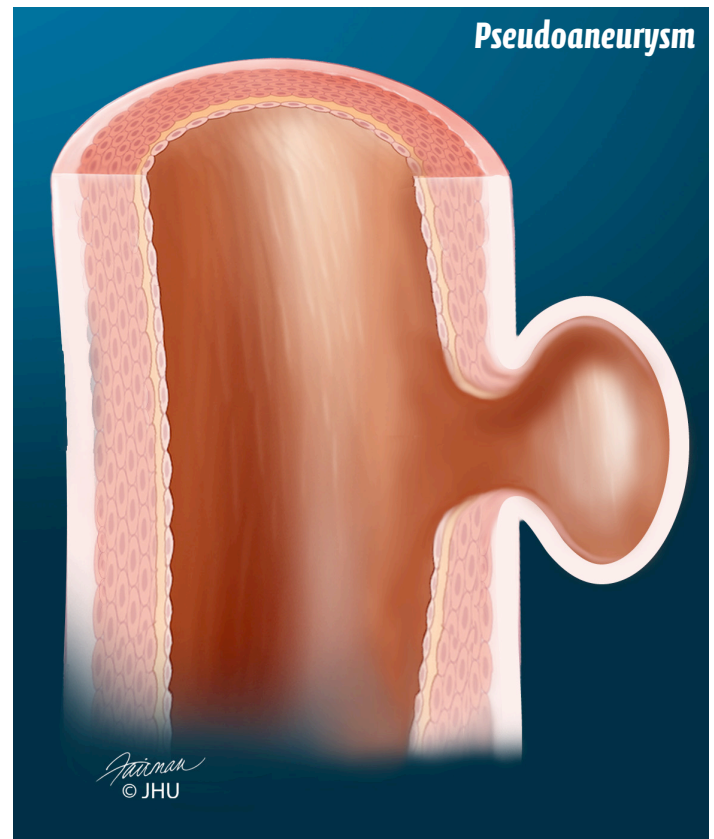
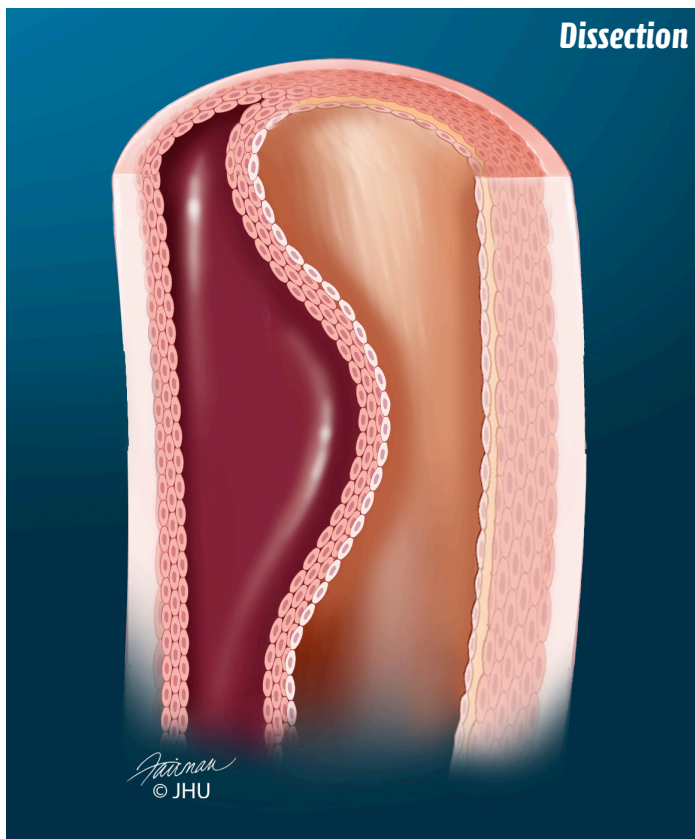
An arterial dissection refers to a tear in the innermost layer, the intima. The presence of a tear leads to a breach in the usual configuration of the three layers of the blood vessel wall. The disruption of the normally smooth intimal surface leads to a cleavage plane, and now there are two passageways for the flow of the blood. The passageway with all three layers intact is called the true lumen, and the passageway outside the tear only has two layers of the blood vessel wall intact, the media and the adventitia.

Aortic dissections are occasionally asymptomatic. More commonly, they cause an array of symptoms

and the manifestation of these symptoms depends on the anatomic location and extent of the tear in the blood vessel wall. A dissection may compromise blood flow to the extremities or important internal organs such as the kidneys, intestines, liver, or spleen. They may also cause pain. Finally, with time the weakened blood vessel wall may expand so that the prior dissection which was once normal in diameter dilates to become a dissection with an aneurysm component as well.

PSEUDOANEURYSM

Pseudoaneurysm literally means “false” aneurysm. Pseudoaneurysms are essentially a contained rupture of a blood vessel. In the area of a pseudoaneurysm, all three layers of the blood vessel wall are disrupted and absent. Therefore, pulsatile blood tracks into the space outside a blood vessel, but surrounding hematoma and tissue contain the blood flow so that free hemorrhage does not typically occur. However, the most dreaded complication of pseudoaneurysm formation is still free rupture of the artery and life threatening bleeding.



SURVEILLANCE

ECHOCARDIOGRAPHY

Echocardiography is a fancy word for ultrasound of the heart and the aorta as it exits the heart. With this test a special gel is applied to the chest area to aid with the conduction of sound waves as the test is being performed. Images of the heart and aorta are transmitted on a special monitor, and cardiologists are then able to interpret these images. Many centers advocate that patients with EDS undergo routine echocardiography in order to evaluate the diameter of the aorta as it exits the heart. This region of the aorta is especially susceptible to aneurysm and dissection formation, and therefore close surveillance with this test is intended to pick up early evidence of aneurysm to prevent the dreaded complication of free rupture. Experts in managing EDS recommend yearly to every three years examination with echocardiography.

CT/MRA

CT stands for Computerized Tomography (CAT scan), and MRA is an abbreviation for Magnetic Resonance Angiography. Both these tests require the patient to lie flat for a period of time in the scanning machine. Both are excellent tests for imaging the blood vessels and other organs within the body. With CT the patient needs to lie still for approximately fifteen minutes, whereas with MRA the patient needs to lie still for longer, approximately thirty to forty-five minutes. In addition, patients who have metal in their bodies may not be able to undergo MRA. Once the diagnosis of EDS has been made, the consensus recommendation is that patients undergo CT or MRA of the entire head and neck, chest, abdomen, and pelvis in order to survey the majority of the vasculature. If the scan is

entirely normal, most experts recommend the next scan occur in approximately three years, in order to minimize radiation exposure. If abnormalities in blood vessels are detected, the CT or MRA will provide insight into the diameter of the blood vessel and also the extent of involvement of the affected blood vessel. These imaging tests will help guide any potential necessary interventions.

ULTRASOUND

Ultrasound is a radiology study that uses a special frequency of sound waves to identify blood vessels and other organs within the body. It does not create images as clear-cut as CT or MRA, however it is non-invasive, relatively inexpensive, can produce reliable blood vessel measurements, and does not expose the patient to radiation. Therefore, it is recommended that patients with EDS undergo annual ultrasound of the carotid arteries and the abdomen to assess the aorta and branching vessels.

TREATMENT INDICATIONS

MEDICAL THERAPY

There is no medication that can cure or prevent the vascular complications associated with EDS. However, there is an important focus on blood pressure management for patients with this disorder. The target systolic blood pressure (top number) should be less than 130. Additionally, supplemental Vitamin C has been recommended to patients with EDS because of its role in the formation of strong connective tissue. Vitamin C theoretically may prevent some of the bruising complications that occur in EDS, but does not prevent or cure the formation of aneurysms, dissections, or pseudoaneurysms. In addition, EDS patients should avoid contact sports as this poses risk to solid organ and blood vessel rupture.

SURGICAL THERAPY

Historically, surgery thought to be so risky that it

was reserved for patients with a catastrophic, life threatening rupture of an organ or major blood vessel. It was thought that patients with EDS had frail tissue and the risks of elective surgery did not outweigh the potential benefits. This can be unsettling to the relationship between physician and patient, because many EDS patients have known aneurysms, dissections, or pseudoaneurysms, which all represent a risk of rupture or other more dangerous complications.

However, there are surgeons who have developed particular experience treating patients with EDS, and certain patients may stand to benefit from an early, elective operation to correct the vascular complications. This is true especially for patients who have known aneurysms, dissections, or pseudoaneurysms that have undergone recent enlargement. If a patient has undergone surgery for other reasons in the past, and there was a history of good quality tissue handling, that particular EDS patient will likely tolerate surgery well.

In the event an EDS patient requires a special catheter known as a central venous line, that catheter should be placed under ultrasound guidance to minimize the risks of hematoma formation and vessel injury. In addition, availability of blood products should be readily available, as the need for blood transfusion is highly likely in patients undergoing repair of vascular complications.

TREATMENT OPTIONS

SURGICAL RECONSTRUCTION

Surgery is undertaken either in an elective fashion after careful consideration, or in the face of imminent life threatening hemorrhage. When a patient develops bleeding from a ruptured artery, one approach can be open surgery with ligation of the

blood vessel. This is a reasonable option when the artery can be sacrificed with no significant negative impact on the patient. Another option would be to repair the artery with a synthetic tube graft (made out of a durable material known as Dacron or Gore-Tex). When doing so, all suture lines should be reinforced with a felt buttress.

ENDOVASCULAR THERAPY

Endovascular therapies are also reasonable options in appropriate patients. Angiography is a procedure whereby the operator places a needle in an artery in the groin and then is able to advance special wires and catheters into the various arteries. With these catheters, special coils can be placed within bleeding blood vessels, and these coils cause a bleeding vessel to thrombose, which means to clot. This procedure is known as coil embolization, and can be a very effective maneuver in patients with pseudoaneurysms, or ruptured aneurysms. Coil embolization can typically only be applied to rupture of smaller arteries, and would not be a reasonable option in a patient with enlargement or an aneurysm of the aorta or its major branches. When angiography is performed, the surgeon should carefully close the access site in the artery with suture, as these sites are prone to later develop vascular complications such as dissection or pseudoaneurysm. Compared with traditional surgery, stent grafts have become an accepted less invasive treatment for aneurysms. However, in general stent grafts are not recommended for patients with EDS, because there is concern that stent grafts are too stiff and may cause further harm to the fragile tissue.

Elective surgical repair can be achieved safely and with acceptable long term results in patients who are appropriately selected to undergo surgery. The decision to proceed with surgery should be made carefully, and after counseling with a vascular surgeon who is experienced in treating EDS patients.