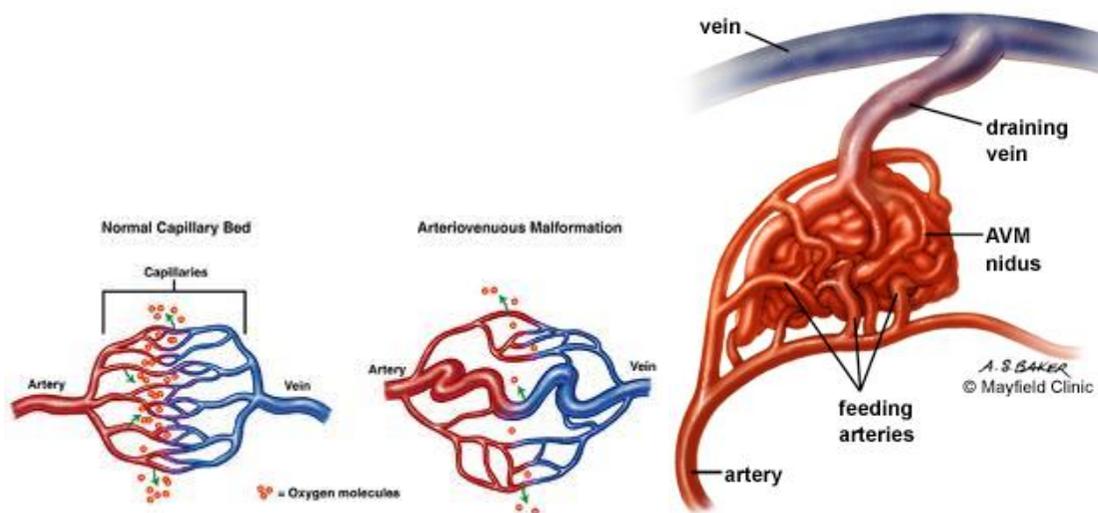


AVMs

Arteriovenous malformations (AVMs) are defects of the circulatory system that are generally believed to arise during embryonic or fetal development or soon after birth. They are comprised of snarled tangles of arteries and veins. Arteries carry oxygen-rich blood away from the heart to the body's cells; veins return oxygen-depleted blood to the lungs and heart. The absence of capillaries—small blood vessels that connect arteries to veins—creates a short-cut for blood to pass directly from arteries to veins. The presence of an AVM disrupts this vital cyclical process. Although AVMs can develop in many different sites, those located in the brain or spinal cord—the two parts of the central nervous system—can have especially widespread effects on the body.



Introduction

Arteriovenous malformations (AVM) are a problem of the vascular (blood vessel) system. AVM result from an abnormal connection between the arteries and the veins and can occur anywhere in the body.

Normally blood flows from arteries to veins through a network of tiny vessels in the tissues known as capillaries. This capillary bed allows oxygen and nutrients to pass from the blood into the tissues and for waste products to be removed. In an AVM, the capillaries are abnormally dilated as shown in the picture below. Sometimes there is no capillary bed at all (fistula).

These abnormal connections allow blood to pass more rapidly from the arteries to the veins (called arterial-venous shunting) with a number of consequences.

- The arteries and veins enlarge and can rupture and bleed.
- The rapid flow can steal blood from surrounding tissues or even cause the heart to fail in very young children.

- Increased pressure in the veins draining the AVM results in congestion of the surrounding tissues which stop working properly Arteriovenous malformations.
- The structural blood vessel abnormality is normally present from birth, although it may not be apparent until later.
- There is no evidence that any foods, activities or medications taken during pregnancy can lead to the development of arteriovenous malformations in the foetus.
- The clinical effect of an AVM will depend on many factors including its size and location and how rapidly the blood is flowing through it.
- Some AVMs are small and never cause a problem, others can be massive and dangerous if not treated.

The vascular system in more detail

Arteries normally carry blood rich in oxygen away from the heart. The oxygen is delivered to the tissues (muscles, brain, liver, skin etc) from the blood via the very smallest vessels known as capillaries. After the blood has passed through the capillaries the blood is brought back to the lungs by the veins. The blood is 're-charged' with oxygen in the lungs, then the oxygen-rich blood is passed to the heart to be pumped again through the arteries to the tissues.

In addition to allowing transfer of oxygen to the tissues, capillaries carry away carbon dioxide, which is a waste product constantly produced by the tissues.

Capillaries also help to exert a dampening effect on the high pressure within the arteries. In their absence this pressure is transferred directly to the veins, which have thinner walls and are used to working at lower pressure. Veins become distorted and damaged by the constant exposure to these abnormally high pressures. They can rupture and bleed. Also, they become congested and cannot drain blood from the surrounding structures properly. Because capillaries are abnormal or lacking in an arteriovenous malformation the normal process of delivery of oxygen is not possible. This may lead to insufficient oxygen delivery to the tissues close to the arteriovenous malformation, despite the high blood flow.

The increased rate of blood flow through an arteriovenous malformation can lead to increased work for the heart. In very young children, occasionally this 'short-circuit' can lead to heart failure requiring urgent treatment.

Signs and Symptoms of AVMs

The effects of arteriovenous malformations are extremely varied and will depend on the size of the malformation, the tissues it involves and the rate of blood flow. Possible complications of arteriovenous malformations include pain and bleeding. Some arteriovenous malformations may produce no symptoms at all, and are only discovered by chance. AVMs confined to the skin may cause cosmetic problems, but do not usually present a significant danger, whereas even a small AVM in the brain can be very serious if it bleeds.

AVMs involving the skin

Although arteriovenous malformations develop before birth, they may not be evident at birth. Sometimes an arteriovenous malformation involving the skin may be seen as a dull red stain at birth or shortly after. The affected area may enlarge and become evident as a swelling, which may feel hot. It is often possible to feel pulsation within the swelling. As the rate of blood flow within the arteriovenous malformation increases over time, there may be darkening of the skin with more pulsation and local warmth.

Early in childhood the arteriovenous malformations that cause staining of the skin can be mistaken for abnormalities called port-wine stains or hemangiomas. Although usually localized to the skin layers, occasionally an AVM seen in the skin can indicate a much larger lesion involving underlying muscle, bone and other organs.

Most arteriovenous malformations involving the brain or internal organs are 'invisible' – ie they do not have any visible mark on the skin.

Associated Problems

Arteriovenous malformations can occur in combination with other disorders including hereditary haemorrhagic telangiectasia (HHT), Parkes Weber syndrome (arteriovenous malformations associated with enlargement of a limb) Wyburn-Mason syndrome (arteriovenous malformations of the brain associated with an abnormality of the retina in the eye), and Cobb syndrome (vascular malformations of the skin with an arteriovenous malformation of the spinal cord).

Investigations

Arteriovenous malformations involving skin and underlying tissues can often be diagnosed clinically. The skin can feel warmer than unaffected areas, and it may be possible to feel a thrill (this is a 'buzzing' feeling, rather like the sensation you get if you touch a purring cat). Using a stethoscope it may be possible to hear a distinctive sound, known as a bruit.. More detailed investigations may then be requested by your doctor. These may include ultrasound, MRI and arteriography.

Treatment

Treatment will depend on the location and size of the arteriovenous malformation and the type of problems that it is causing. Each case needs to be assessed on an individual basis. The expertise of a number of specialists may be necessary, depending on the location of the lesion. In some cases, the blood flow through the AVM can be decreased using techniques to block the blood vessels. This procedure is called embolization and is performed by an interventional radiologist. It is very rare that embolization offers a 'cure' but it can be helpful in making the AVM less troublesome. *Written by Glover M, Barnacle A, Robertson F. Great Ormond Street Hospital 3.9.1*