A Novel Approach to Using Echo in the Assessment of Arterio-Venous Malformations

LIZ HODSON,[1] *CHIEF CARDIAC PHYSIOLOGIST*
WILL NICOLSON,[1] *CONSULTANT CARDIOLOGIST*
PROFESSOR MATTHEW BOWN,[1] *CONSULTANT VASCULAR SURGEON*
[1]GLENFIELD HOSPITAL.


Liz Hodson


Dr Will Nicolson


Professor Matthew Bown

Arteriovenous malformations (AVMs) are abnormal, entangled blood vessels that make non uniform communications between arteries and veins. These communications can create a low resistance shunt which bypasses the capillaries and tissue in an area of the body and can cause tissue damage and pain to the affected site. AVMs can occur anywhere in the body. High flow AVMs, where there is a significant volume of fast flowing blood travelling directly from the arterial circulation into the venous circulation require the heart to work harder in order to deliver normal blood flow to other areas. This process is similar to that of intra-cardiac and other left-to-right shunts. Although individuals with AVMs will often have these present at birth, they may not be discovered until later in life. As the body grows and/or hormonal changes take place, AVMs tend to increase in size as they are hormone sensitive. Some AVMs may not become apparent until puberty, pregnancy or following trauma or injury to the site.

The Schobinger Staging System is used as a scale to grade the AVM and how it changes over time-

- **Stage I** (Quiescence): The AVM is stable, however the skin on top of the AVM may be warm and discoloured
- **Stage II** (Expansion): The AVM gets larger. A pulse can be heard or felt in the AVM
- **Stage III** (Destruction): The AVM causes pain, bleeding or functional issues
- **Stage IV** (Decompensation): High output cardiac failure occurs

The effect of high flow AVMs on the cardiac status is not widely reported; however, it is well known that patients with significantly shunting AVMs may experience high output cardiac failure. This type of failure can occur at any point in the presentation of an AVM. Some studies have documented that heart failure is one of the initial presenting symptoms in AVM. As we know, congestive heart failure is characterised by low cardiac output with reduced left ventricular function, however high output cardiac failure is defined as cardiac output of more than 8 L/min or a cardiac index of greater than 3.9 L/min/m.^{2} High output heart failure is characterised by a reduction in systemic vascular resistance (e.g. due to a large low resistance AVM shunt). The heart is unable to increase output sufficiently to maintain blood pressure and volume for tissue perfusion and this in turn activates the renin-angiotensin-aldosterone system, thus causing salt and water retention and the fluid overload clinical features of heart failure. One of the most obvious physical findings in high output heart failure patients is the presence of warm peripheries rather than the cold ones found in low cardiac output states. This occurs due to low systemic vascular resistance and vasodilatation.

For patients with AVMs affecting the lower and upper limbs, we have devised an echo protocol at our hospital to assess the effect of the AVM on cardiac output. This has been developed in people with high flow AVMs that have reached Stage III or IV of the Schobinger scale and may be showing signs of extreme discomfort in the affected limb or clinical features of high output cardiac failure.

Our protocol has been written by Dr Will Nicolson (Consultant Cardiologist) and Professor Matthew Bown (Consultant Vascular Surgeon). The purpose of the transthoracic echo is to first assess cardiac structure and function and secondly to quantify the effect of excluding the AVM from the circulation on cardiac output (i.e. removing the arteriovenous shunt). The echo can provide important evidence to inform a decision as to whether to operate on the AVM or not. Although AVMs can occur almost anywhere in the body, for the purposes of using this method of echocardiography, assessment using our cuffed and un-cuffed protocol can only be made for AVMs present in the arms and legs where a blood pressure cuff can be applied to the limb above the AVM.

AVM Protocol Echo

1. The process involves performing a standard BSE dataset echo and measuring the following-

- Heart rate
- LVEF
- LVOT Diameter
- LVOT VTI
- Calculate stroke volume and cardiac output

2. Using a manual blood pressure monitor the BP should be checked on the arm or leg not affected by the AVM and recorded. The cuff should then be inflated on the non-affected contralateral limb to 10mmHg above the systolic BP.

3. Wait for 30 seconds, keeping the cuff constantly inflated to 10mmHg above the systolic blood pressure. Re-take the images to be able to measure heart rate, LVEF, LVOT VTI, stroke volume and cardiac output again.

4. Check the blood pressure on the arm or leg affected by the AVM and record it.

5. Inflate BP cuff 10mmHg above systolic blood pressure in the limb affected by the AVM. Wait 30 seconds and measure heart rate, LVEF, LVOT VTI, stroke volume and cardiac output.

The blood pressure cuff should not be left inflated for more than 2 minutes. If necessary, deflate the cuff for 1 minute and then re-inflate. If the patient develops pain in the arm/leg, deflate the cuff and abandon this part of the echocardiogram.

Where a leg AVM is being assessed a vascular tourniquet is often required and a doctor or nurse will need to be in attendance to apply and inflate the vascular tourniquet.

During an AVM protocol echo we expect to see a decrease in cardiac output when we block flow in the AVM with the blood pressure cuff. Inflating the blood pressure cuff could have an impact on cardiac output through other mechanisms i.e. pain induced anxiety increasing heart rate and therefore cardiac output. Due to this, the contralateral arm / leg should be assessed first. By analysing the echo data we are able to ascertain if the arm or leg AVM is having a significant impact on the patient's cardiac status. The findings of the echo, along with the symptoms and clinical findings of each patient work towards a treatment plan. For some patients, if the extent of the effect on cardiac output is great and symptomatic burden is high from both a cardiac and arm/leg pain perspective, the only form of curative treatment is amputation of the affected limb. With this extreme treatment in mind, extensive testing in addition to echo is also required; this includes right heart catheterisation and cardio-pulmonary exercise testing.

As we build up the numbers of patients with AVM where echo is used in their assessment, there is the potential to adapt the protocol where necessary. Currently there is scope to increase the blood pressure margin of the cuff to 50mmHg for arm and 70mmHg for leg AVMs to assess if this more clearly identifies the significance of the AVM on the cardiac status. Once used in practice, the two methods will be compared.

Currently there is only a small cohort of patients that have undergone our AVM protocol echo. We hope that at some point in the future these studies will provide pilot data to justify a study to more accurately assess the role of transthoracic echocardiography in assessment of the cardiac status of patients with arm and leg AVMs.

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