

2020 PAVM and HHT Research Newsletter

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<https://www.imperial.nhs.uk/our-services/respiratory-medicine/hht-pavms>

PULMONARY ARTERIOVENOUS MALFORMATIONS (PAVMs)

MRI SCANS- SAFE AFTER EMBOLISATION OF PULMONARY AVMS:

In 2019 the Hammersmith team published this important paper. Please bring this to the attention of your doctors and radiologists to avoid unnecessary MR scan delays:

ALSAFI A, JACKSON JE, FATANIA G, PATEL MC, GLOVER A, SHOVLIN CL. Br J Radiol 2019;92:20180752. PATIENTS WITH IN-SITU METALLIC COILS AND AMPLATZER VASCULAR PLUGS USED TO TREAT PULMONARY ARTERIOVENOUS MALFORMATIONS SINCE 1984 CAN SAFELY UNDERGO MAGNETIC RESONANCE IMAGING.

WHY DO I NOW FEEL BREATHLESS?

If you become breathless, most doctors think that PAVMs have got worse. But this is not usually the case. Our 2019 respiratory papers show two much more common reasons: Less air getting in and out of your lungs (even a slight change as in asthma or COPD), or Developing anaemia/ low haemoglobin. Please let your doctors know, so they can check:

GAWECKI F, MYERS J, SHOVLIN CL. BMJ OPEN RESPIR RES. 2019 MAR 1;6(1):e000351.
GAWECKI ET AL. QJM. 2019 MAY 1;112(5):335-342. DOI: 10.1093/QJMED/HCZ023.
EXERCISE CAPACITY REFLECTS AIRFLOW LIMITATION RATHER THAN HYPOXAEMIA IN PATIENTS WITH PAVMS.

WHY DO YOU TELL US TO USE ANTIBIOTICS BEFORE DENTAL CARE AND SURGERY?

Whether or not PAVMs are due to HHT, the European Reference Network VASCERN generated a summary manuscript which your doctors and dentists may wish to read:

SHOVLIN C, BAMFORD K, SABBÀ C, MAGER HJ, KJELDSSEN A, DROEGE F, BUSCARINI E, DUPUIS-GIROD S; VASCERN HHT. HAEMATOLOGICA. 2019 FEB;104(2):E85-E86.
PREVENTION OF SERIOUS INFECTIONS IN HEREDITARY HEMORRHAGIC TELANGIECTASIA: ROLES FOR PROPHYLACTIC ANTIBIOTICS, THE PULMONARY CAPILLARIES-BUT NOT VACCINATION.

VASCERN HHT also generated COVID statements for people with AVMS, including pulmonary AVMS: see <https://vascern.eu/home/vascerns-covid19-recommendations/>:

HEREDITARY HAEMORRHAGIC TELANGIECTASIA (HHT)

SEVERE ANAEMIA IN HHT

If you are in the group of people that seem to need more iron or blood transfusions, than you expect from your amount of bleeding, do ask your doctors to look at:

THIELEMANS L, LAYTON M, SHOVLIN CL. HAEMATOLOGICA 2019 APR;104(4):E127-E130
LOW SERUM HAPTOGLOBIN AND BLOOD FILMS SUGGEST INTRAVASCULAR HEMOLYSIS CONTRIBUTES TO SEVERE ANEMIA IN HEREDITARY HEMORRHAGIC TELANGIECTASIA.

CAN I TAKE BLOOD THINNERS IF I HAVE HHT, AND WHAT DO YOU RECOMMEND?

We know that blood thinners often make little difference to HHT nosebleeds, but can make nosebleeds worse - everyone is different. Looking at patients across Europe, the European Reference Network VASCERN showed that the exact type of blood thinner also makes a difference, so if you need blood thinners, please ask your doctors to read:

SHOVLIN CL, MILLAR CM, DROEGE F, KJELDSSEN A, MANFREDI G, SUPPRESSA P, UGOLINI S, COOTE N, FIALLA AD, GEISTHOFF U, LENATO GM, MAGER HJ, PAGELLA F, POST MC, SABBÀ C, SURE U, TORRING PM, DUPUIS-GIROD S, BUSCARINI E; VASCERN-HHT. ORPHANET J RARE DIS. 2019 AUG;14(1):210.
SAFETY OF DIRECT ORAL ANTICOAGULANTS IN PATIENTS WITH HEREDITARY HEMORRHAGIC TELANGIECTASIA.

THE EUROPEAN REFERENCE NETWORK VASCERN HHT have also published many additional statements and papers for people with HHT, and are summarising in a single paper for 2021. In the meantime, please check the VASCERN HHT website at <https://vascern.eu/expertise/rare-diseases-wgs/hht-wg/> for:

- COVID Statements,
- Do's and Don't in Common Situations,
- and other topics including our YouTube video "HHT from VASCERN HHT".

Professor Claire Shovlin, December 2020

STOP PRESS: 2021 UPDATES:

1. We are really sorry that our clinical services were so delayed by COVID, but are pleased to say that after a year of nearly all teleconsultations, we will be starting to resume face to face services from April 2021.
2. Brexit has had major effects on our roles in Europe, but after 5 years, the wisdom of European experts in VASCERN is just as important for us in the UK, and they have also indicated they want to still be working with us, to help deliver the best care for people with HHT and AVMS. Please watch this space.

3. 2021 SURVEY ON HHT AND PAVMs.

As you know, we have conducted many research studies to look into different medical problems that people with HHT and PAVMs experience, in order to provide the communities with better advice about their lifestyle and health.

We invite you to help us with our 2021 research, by filling out a short confidential survey. This has been reviewed by the NHS HRA London - South East Research Ethics Committee, and should take about 15-20 minutes to complete.

You can find out more information, and complete the survey in your own time at:

<https://eu.research.net/r/Imperial2021HHTPAVMSurvey>