



VASCERN HHT Statement on COVID-19

A statement from the European Reference Network for Rare Multisystemic Vascular Diseases (VASCERN) for people with hereditary haemorrhagic telangiectasia (HHT) and their doctors:

- 1. People with HHT should follow the standard Public Health Measures as recommended in their specific country.**
 - 1.1. These are directed at reducing the spread of infection, and strategies differ slightly between countries.
- 2. People with HHT should be no more and no less concerned about COVID-19 than the general population without HHT.**
 - 2.1. There is no reason to think people with HHT [1] will be at higher or lower risk of infection [2,3], or complications if they become infected.
 - 2.2. Some people with HHT, as for some people in the general population, may be less able to tolerate the extra demands placed on their bodies if they suffer infection with complications, but this will be a small group and should not be applied to all HHT patients.
- 3. The presence of HHT or AVMs in someone who currently has a normal or high exercise tolerance should not limit their access to medical treatment compared to someone without HHT or AVMs of the same age.**
 - 3.1. People with HHT have normal life expectancy managed in Europe, [4,5] likely attributed to the beneficial effects of reduced cancer, reduced rates of cancer [4,6,7] and fewer heart attacks [8]
 - 3.2. Anyone with normal or high exercise tolerance (able to walk uphill quite briskly without stopping) will have good cardiorespiratory reserve [9,10].
- 4. For those who are obliged to self-isolate because of the general situation**
 - 4.1. Maintain normal treatment regimes if possible. **It is very important that iron supplements for anaemia are continued.**
 - 4.2. Avoid sedentary states- exercise is recommended as discussed further for the general population [11]

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During the current epidemic, reference centres may be unable to perform elective diagnostic/therapeutic procedures for people with HHT but they continue to assist urgent/emergency presentations

Further reading if required:

General HHT:

[1] VASCERN HHT 2019 Orphanet Definition of HHT https://www.orpha.net/consor/cgi-bin/OC_Exp.php?Expert=774

Infection/Viral infection

[2] VASCERN HHT Immunity-and-Inflammation-Workshop <https://vascern.eu/wp-content/uploads/2018/03/Immunity-and-Inflammation-Workshop-2017.pdf>

[3] VASCERN HHT Prophylactic antibiotics, the pulmonary capillaries, and vaccination in HHT <http://www.haematologica.org/content/104/2/e85.long/>

Survival/life expectancy:

[4] Kjeldsen A, Aagaard KS, Tørring PM, Möller S, Green A. 20-year follow-up study of Danish HHT patients-survival and causes of death. Orphanet J Rare Dis. 2016 Nov 22;11(1):157 <https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5120428/>

[5] HHT from VASCERN HHT An Introduction to hereditary haemorrhagic telangiectasia <https://www.youtube.com/watch?v=0YjWf7Agn40&feature=youtu.be>

[6] Hosman AE, Devlin HL, Silva BM, Shovlin CL. Specific cancer rates may differ in patients with hereditary haemorrhagic telangiectasia compared to controls. Orphanet J Rare Dis. 2013 Dec 20;8:195. <https://orjrd.biomedcentral.com/articles/10.1186/1750-1172-8-195>

[7] Duarte CW, Murray K, Lucas FL, Fairfield K, Miller H, Brooks P, Vary CPH. Improved survival outcomes in cancer patients with hereditary hemorrhagic telangiectasia. Cancer Epidemiol Biomarkers Prev. 2014 Jan;23(1):117-125. <https://cebp.aacrjournals.org/content/23/1/117.long>

[8] Shovlin CL, Awan I, Cahilog Z, Abdulla FN, Guttmacher AE. Reported cardiac phenotypes in hereditary hemorrhagic telangiectasia. Int J Cardiol. 2016 Jul 15;215:179-85. [https://www.internationaljournalofcardiology.com/article/S0167-5273\(16\)30679-9/fulltext](https://www.internationaljournalofcardiology.com/article/S0167-5273(16)30679-9/fulltext)

Exercise, compensations and conditioning - with specific reference to normal capacity despite hypoxaemia from pulmonary AVMs

[9] Gawecki F, Myers J, Shovlin CL. Veterans Specific Activity Questionnaire (VSAQ): a new and efficient method of assessing exercise capacity in patients with pulmonary arteriovenous malformations. BMJ Open Respir Res. 2019 Mar 1;6(1):e000351 <https://www.ncbi.nlm.nih.gov/pmc/articles/PMC6424292/>

[10] The British Thoracic Society Clinical Statement on Pulmonary AVMs <https://www.brit-thoracic.org.uk/quality-improvement/clinical-statements/pavm/>

[11] Shovlin CL, Moorthy K, Lees C. Covid-19: Home based exercise activities could help during self isolation BMJ 2020 <https://blogs.bmj.com/bmj/2020/03/16/covid-19-home-based-exercise-activities-could-help-during-self-isolation/>