

2017 Research Newsletter: Hereditary Haemorrhagic Telangiectasia (HHT)

Claire L. Shovlin, PhD FRCP

Imperial College London, and HHTIC London, Hammersmith Hospital, Imperial College Healthcare NHS Trust, London, UK

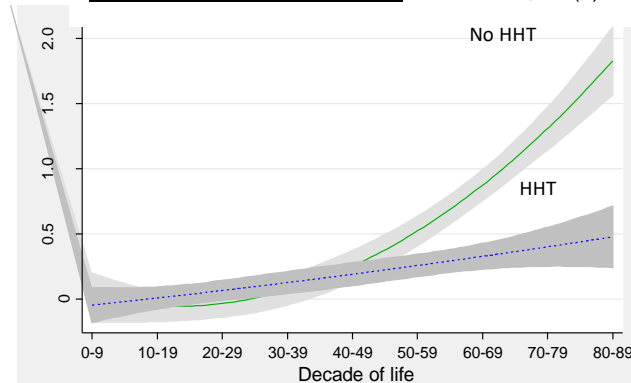
We would like to thank all of you who have helped us with our recent research studies. Here we provide information to summarise important new findings. You and your doctors can find further information, and access to most of the papers, via <https://www.imperial.ac.uk/people/c.shovlin/page/hht-and-pavms.html>

HEALTH BENEFITS FROM HHT

Why do some HHT families report 'people with nosebleeds seem to live longer?'

We think we now have two parts of the answer. First, it seems that people with HHT have lower rates of certain cancers. This was shown by us in a survey that many of you completed, and has since been confirmed by a separate study in Denmark. You may be surprised, but some new cancer treatments are effectively giving people HHT, using an antibody instead of an inherited gene mutation.

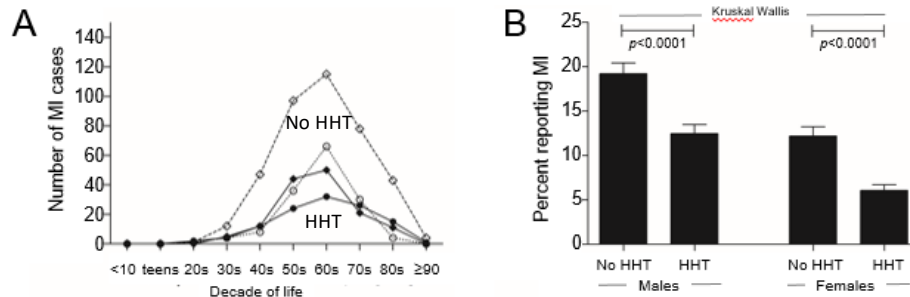
HOSMAN ET AL, ORPHANET J RARE DIS 2013 Feb 28;368(9):876-8.



Although some cancers occurred at equal rates in people with and without HHT, overall, there were fewer cancers in HHT. This was best shown for lung cancer (see graph). Fewer cases were seen in people who had HHT, despite them smoking as much, if not more, than people without HHT

Secondly, it seems people with HHT have fewer heart attacks ("MIs"), even though they are often anaemic and have other medical issues that might make you think it would put a strain on the heart. This has been suspected for many years- some heart doctors have even told some of you that "People with HHT have strong hearts"- and they are right. Heart attacks did occur- at about the same age as in the general population (see A)- but in our study, people with HHT had fewer (B)

SHOVLIN ET AL, INT J CARDIOL 2017 Jul 15;215:179-85



HHT NOSEBLEEDS:

Why noses may bleed at certain times for people with HHT:

We have really never understood why people with HHT have their nosebleeds at particular times. The patterns that you reported to us suggest that for some of you, the foods you eat might influence nosebleeds. Two possible reasons are that 1) some foods contain natural blood thinners, and 2) some foods make blood vessels squeeze close and open again (triggering migraines in many people). So we suggest if you have a particularly bad nosebleed, think about what you have eaten in the previous few hours, and see if you can identify any patterns in your "nosebleed food diary". We have some evidence to suggest some of you already make slight changes to your diet.

SILVA ET AL, LARYNGOSCOPE 2013 May;123(5):1092-9

ELPHICK & SHOVLIN, LARYNGOSCOPE 2014 Jul;124(7):1521-8

FINNAMORE ET AL, ORPHANET J RARE DIS 2017 IN PRESS

Blood thinners and HHT

There is no doubt that for some of you, taking a blood thinner such as aspirin or warfarin can make your nosebleeds worse. Some of you even report this with natural blood thinners such as fish oils. Surprisingly however, nearly half of the people with HHT we studied said blood thinners made no difference to their nosebleeds:

- 1) If you are struggling with your nosebleeds, we suggest you avoid non essential items such as fish oil supplements. We are surprised how many of you take supplements, usually without letting your doctors know! Also, don't use aspirin, or non steroidal painkillers when there are good alternatives that do not thin the blood.
- 2) But if your doctor thinks you should really be given a blood thinner to prevent heart attacks, strokes, blood clots or another medical problem, then there should be a sensible discussion. It may be possible for you to have such blood thinners, and you will not know unless it is tried. Your doctor should know that our data on safety relate to aspirin, warfarin and heparin, and that there seem to be some newer agents (NOACs) that cause more problems. People with HHT who had problems with one form of blood thinning treatment often managed better with others. At the moment, we can't predict what will happen and should have more evidence later in the year

DEVLIN ET AL, NEW ENGL J MED 2013 Feb 28;368(9):876-8

CHAMALI ET AL, INTRACT RARE DIS RES 2016 May;5(2):109-13

STOP PRESS! We have just learned that we and our 7 European Partners have been approved as the HHT arm of a new European Reference Network on Rare Inherited Multisystemic Vascular Diseases- VASCERN (<https://vascern.eu/>). Watch this space!