The ‘Celtic Curse’ – and the 50 Scots found at risk

◆ Haemochromatosis is the most common genetic condition in Scotland with work ongoing to improve prevention of serious illness among patients

Alison Campsie
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Around 50 Scots have received letters informing them they have the gene linked to a disease dubbed the “Celtic Curse” given its high prevalence across Scotland and Ireland.

The new cases follow fresh analysis of the “Viking Genes” database, which holds information on around 10,000 people with at least two grandparents from Orkney, Shetland and the Western Isles.

The database has been used to detect those with the pathogenic genes that cause haemochromatosis – the most common genetic condition in Scotland. While the majority of those in the database live in the island groups, many are known to live in Glasgow, Edinburgh and Aberdeen.

Haemochromatosis causes a build up of iron in the body, which can lead to a range of health issues, from diabetes to liver cancer and heart failure.

Estimates suggest up to one in 100 Scots carry the gene variant, with the research aiming to refine numbers and the location of carriers in a bid to improve screening and increase prevention of serious health issues.

Professor Jim Flett Wilson, from Edinburgh University, who set up the Viking Genes database, has led the research. It follows his breakthrough work on mutations of the BRCA1 and BRCA2 genes, which are linked to a higher incidence of breast and ovarian cancer among women in Orkney, with his findings leading to mass community screening and treatment.

Prof Wilson said: “I became aware that haemochromatosis is known to be very common in Scotland and indeed Ireland, and either Ireland or Scotland is probably the highest in the world.

“These letters only went out three weeks ago. People need to digest the information.

“Some will need treatment, some not but it is better to know. This is part of the idea of preventative medicine rather than waiting until you are broken and harder to fix.”

Those who suffer from haemochromatosis report a range of symptoms, from extreme tiredness to mood swings, brain fog, weakness, joint pain, balance problems and itchy skin. While there is no cure, relatively simple treatment can effectively bring down iron levels, with patients undergoing regular venesections – similar to blood donation. If untreated, a build up of iron in the body damages vital organs over time.

Prof Wilson said the Viking Genes research into cancers had led to “multiple instances“ of people’s lives being saved, with those affected seeking out check-ups and going on to get treatment and then the all clear.

He said: “And they say ‘it is because of your letter that I went and did this’. We predict something similar will be going on in households as people are reading these letters and being checked.”

Prof Wilson said there was a range of theories over why the ‘Celtic Curse’ was so prevalent in Scotland and Ireland. A 4,000-year-old skeleton excavated on Rathlin Island off the coast of Northern Ireland was found to have the gene variant.

Prof Wilson said: “That skeleton was a carrier of haemochromatosis. So it has been in Northern Ireland and Scotland area for 4,000 years bare minimum.”

He added: “I think that everybody in Scotland and in Ireland and England who has this variant, they all descend from a person 4,000, 5,000, 6,000 years ago and everyone is a descendant of this founder individual. That is all that has happened. They haven’t moved all that much and they are all still there.”

Prof Wilson’s discovery of the new cases follows a grant from Haemochromatosis UK, with the funding to produce a nationwide map on cases. A total of 29 regions are being plotted, including nine in Scotland.

Prof Wilson said: “I strongly believe there should be screening, so I saw the chance to do a study to ask the question where in England, Scotland Ireland and Wales is the most common, where are the hot-spots, are there any cold spots. So far it is very vague, so we have looked in 29 regions and we are still finishing it off to make a map.

“When eventually screening does happen when we [anyone] saying ‘let’s have everyone in Scotland or Britain screened’.

persuade the powers that screening would be a good idea as it prevents disease down the line and saves money, there is not going to be

“They are going to have to start somewhere and you should start where there is the most haemochromatosis. So our map, people will be able to use it as a guideline of where should screening be started.

“It is going to be an interesting scientific question, but it has an immediate and direct practical purpose. It is about ensuring that the people of Scotland get good healthcare based on evidence.”

Data from England will come from the DigiTrials bank of more than 63 million patient records.

Neil McClements, chief executive of Haemochromatosis UK, said: “Haemochromatosis is a relatively common condition in Scotland.

“It is massively underdiagnosed, so the work that the charity has been doing – and we are in our 35th year – supports people at risk and those affected by the condition. But we also try and stimulate research to improve clinician and public awareness.

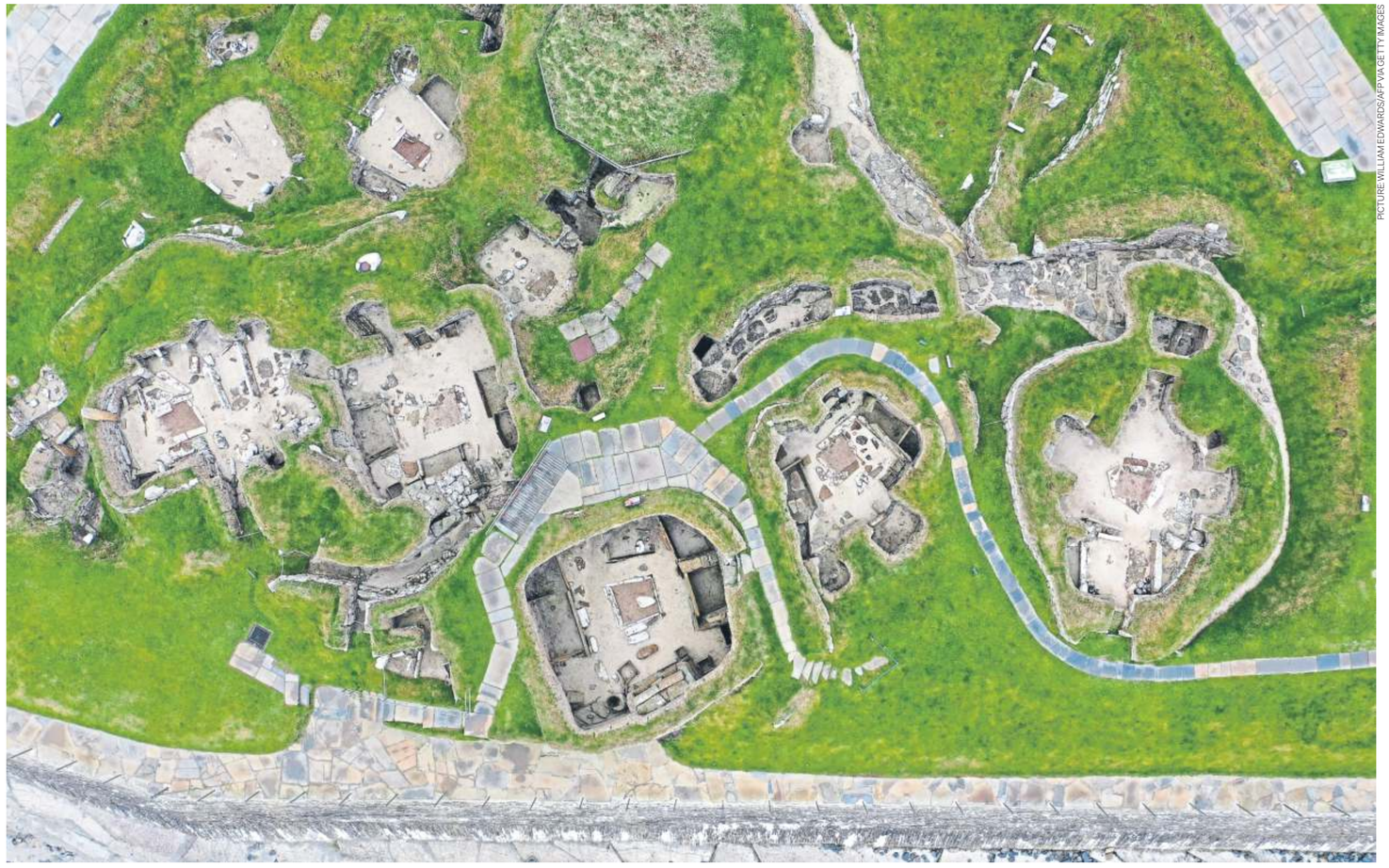
“The map will be incredibly useful. If you are trying to run a centralised healthcare system like the NHS, if you don’t know where your patient groups are it is very hard to allocate resources to the right geographic area.

“In Scotland, there is a kind of belt [of cases] that runs right the way across from Aberdeen to Inverness and right across to the Western Isles. It is particularly prevalent in that kind of arc and there are historical reasons behind that.

“Island communities are close familial communities. So what you end up with is a situation where people tend to be born, live, grow up, maybe travel off the island, come back to the island to have a family and they do that with other people who follow a similar pattern.

“So you end up with a concentration of the gene pool and that is why in the Western Isles it is particularly prevalent.”

Mr McClements said NHS Western Isles was “particularly good” at treating those with haemochromatosis.



PICTURE: WILLIAM EDWARDS/AFP VIA GETTY IMAGES

Main: Skara Brae Neolithic settlement on Orkney. Inset: Miranda McHardy who died from haemochromatosis aged 59. Left: Prof Jim Wilson. Top right: A settlement on Shetland

> “These letters only went out three weeks ago. People need to digest the information”

The estate owner with Celtic Curse who slept so much he was called ‘half man, half mattress’

CASE STUDY

Alison Campsie

Sir George Forbes-Leith suffered such tiredness in the afternoon that a friend started calling him “half-man, half-mattress”.

It was only when his cousin went to the doctor after a long period of “not feeling great” that a diagnosis of haemochromatosis followed.

Sir George, who owns Fyvie Estate in Aberdeenshire, was diagnosed around four years ago. His sister, Miranda McHardy, was also identified as a carrier of the gene variant that causes the condition.

Sir George said: “My cousin was asked if any of her relations had any health problems and she said ‘yes, actually, Miranda’. Miranda had had heart failure, kidney failure, liver failure and she had been ill her whole life.”

Haemochromatosis is an overload of iron in the blood that, if left untreated, can start to attack the organs and cause serious illness.

Normal iron levels in the blood are around 100 but, following testing, it emerged Mrs McHardy’s levels were at 5,000 while her brothers were at around 1,200.

Sir George said: “I was always feeling a bit tired in the afternoons and one chap christened me ‘half man, half mattress’. I could fall asleep every afternoon without any problem. I just presumed it was me getting old.”

Ms McHardy, 59, of Banchory, died from complications from haemochromatosis in 2023.

Sir George continues to be treated for the condition by undergoing venesections, which remove blood from the body to bring down iron levels in a process much like giving blood.

He said the ill health of other family members throughout time started to make sense following diagnosis.

“Genetically, our grandmother must have had it,” he said. “She was in a wheelchair aged 55, but it was put down as rheumatism. My grandfather and father both had heart problems and were always feeling tired and spent most of the time asleep.”

Sir George initially had six months of fortnightly venesections, which removes blood to bring down iron levels, and now undergoes the procedure every six months.

“Once the iron levels went down, the tiredness went away,” he said. “I feel a lot more alert during the day.”

Sir George addd: “The cure is not drugs, the cure is simply becoming a blood donor.”

Since Mrs McHardy’s death, her family and friends have been trying to raise awareness of the condition.

Screening of people in 25,000 households in north-east Scotland was supported in part by money raised by the family for Haemochromatosis UK.

Sir George Forbes-Leith was affected by extreme tiredness