**Table 1**

| GGCX mutation | Age (Age of onset) | Genotype | Non-haemostatic phenotype/s | γ-carboxylated GRP ([%] at 10μM $K_1$) | γ-carboxylated MGP ([%] at 10μM $K_1$) | γ-carboxylated BGLAP ([%] at 10μM $K_1$) | Comments | VKORC1 c.-1639 | Ref. |
|---|---|---|---|---|---|---|---|---|---|
| R83W<br>Q374X | 48 year<br>(27 year) | CH | PXE-like phenotype | 26.2 %<br>- | 31.7 %<br>- | 15.8 %<br>- | | | Li, Schurgers et al., 2009 |
| G537A<br>Q374X | 46 year,<br>44 year | CH | PXE-like phenotype + Atherosclerosis | 18.4 %<br>- | 102 %<br>- | 155.8 %<br>- | | | Vanakker et al., 2007 |
| V255M<br>S300F | 16 year | CH | PXE-like phenotype + Cardiac abnormality | 30.7 %<br>1.5 % | 10.2 %<br>2.9 % | 57.3 %<br>0 % | | | Li, Grange et al., 2009 |
| G558R<br>F299S | 40 year | CH | PXE-like phenotype | 31.7 %<br>0 % | 18.2 %<br>1 % | 65.8 %<br>0 % | | | Vanakker et al., 2007 |
| H404P<br>R485P | 47 year | CH | Mild skin symptom + Calcified peripheral arteries | 34.8 %<br>57.7 % | 8.9 %<br>87.1 % | 0.4 %<br>47.7 % | | GG | Watzka et al., 2014 |
| R476C<br>WT | 67 year<br>(3 year) | HTZ | PXE-like phenotype + Atherosclerosis | 41.7 %<br>100 % | 93.5 %<br>100 % | 56.2 %<br>100 % | Re-evaluation of genotype | | Vanakker et al., 2007 |
| R476H<br>WT | 32 year<br>(18 year) | HTZ | PXE-like phenotype | 95.7 %<br>100 % | 77.9 %<br>100 % | 57.1 %<br>100 % | Re-evaluation of genotype | | Vanakker et al., 2007 |
| R83P<br>R83P | 3 year | HMZ | Facial Dysmorphism + Septal defect | 12.6 %<br>12.6 % | 26.5 %<br>26.5 % | 34 %<br>34 % | Expected to develop PXE-like phenotype | AA | Watzka et al., 2014 |
| L394R<br>L394R | 5 months, newborns | HMZ | - | 33.3 %<br>33.3 % | 36.1 %<br>36.1 % | 0 %<br>0 % | Expected to develop mild skin phenotype | | Brenner et al., 1998 |