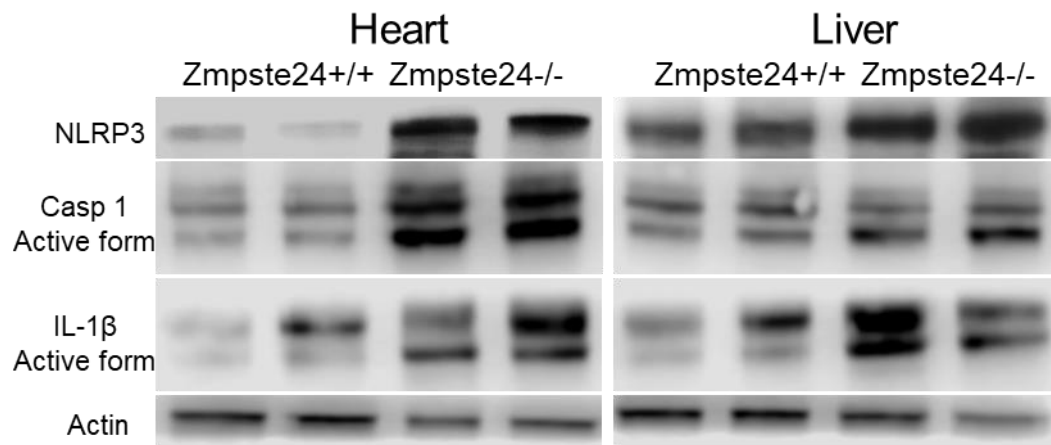
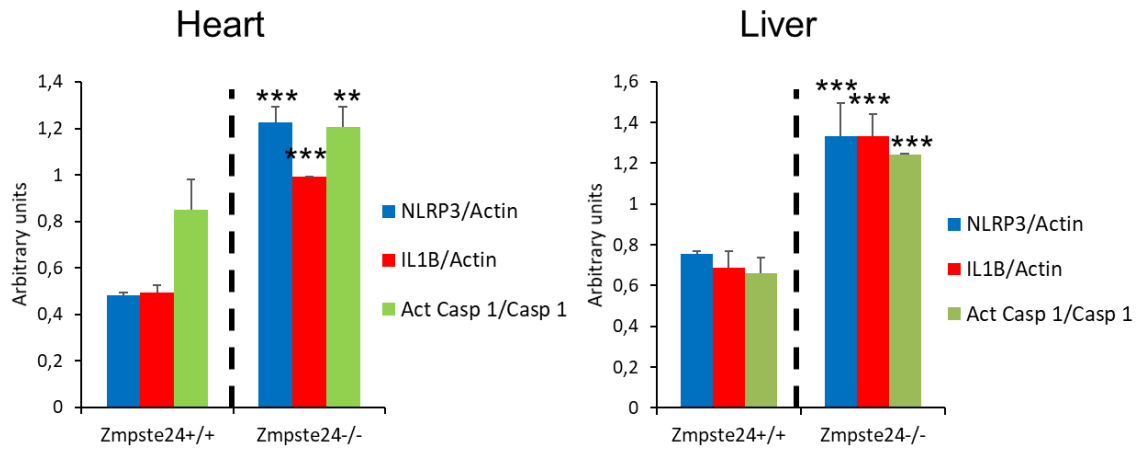


## Appendix

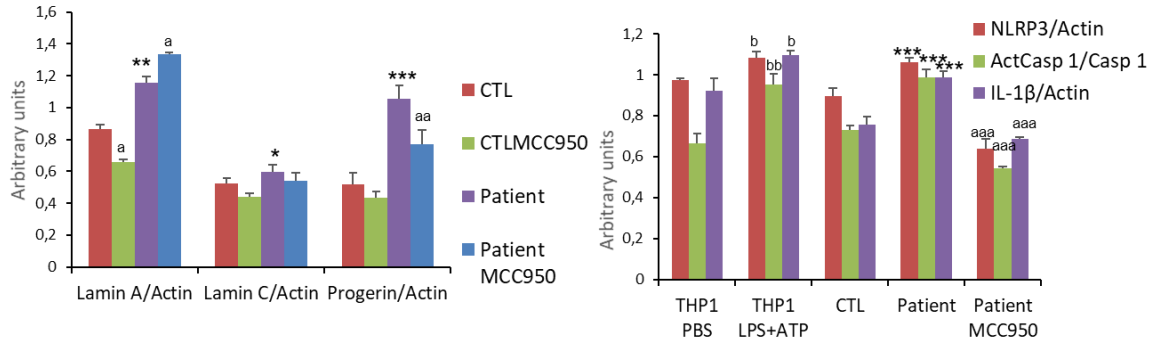
1. Appendix Figure S1. NLRP3-inflammasome complex protein expression with more representative animals.
2. Appendix Figure S2. Densitometry of Figure 1 R2.
3. Appendix Figure S3. Densitometry of Figure 2 R2.
4. Appendix Figure S4. Effect of the MCC950 treatment in skin fibroblasts from HGPS patient 2.



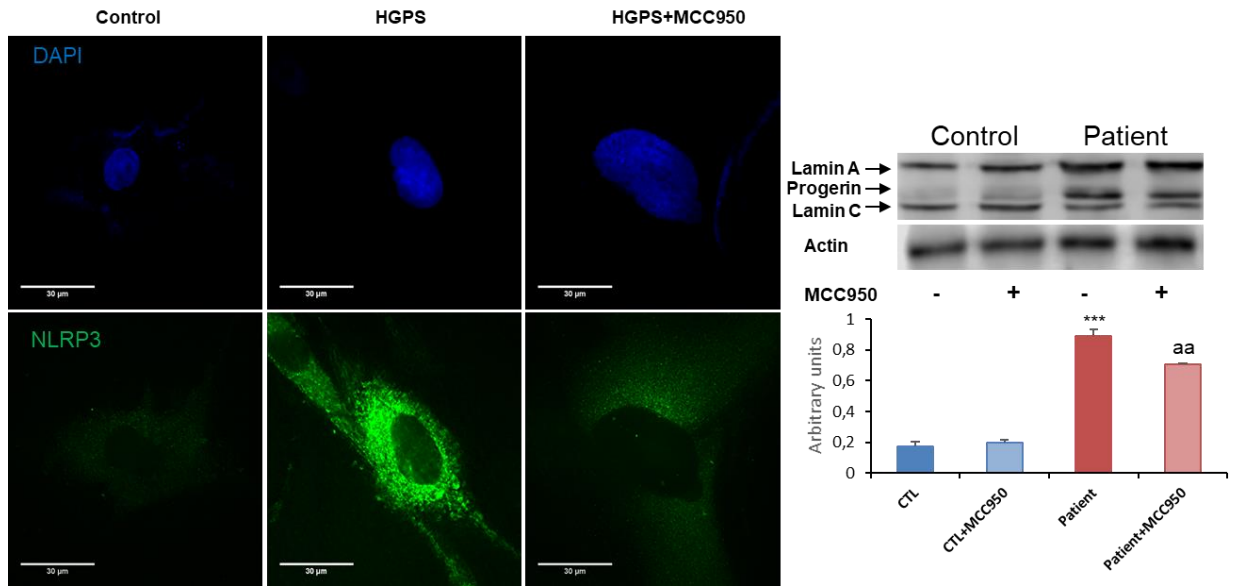
Appendix Figure S1. NLRP3-inflammasome complex protein expression with more representative animals. Western blot analysis with representative blot including NLRP3, caspase 1, IL-1 $\beta$  and actin levels in heart and liver tissues from wild-type and Zmpste24<sup>-/-</sup> mice.



Appendix Figure S2. Densitometry of Figure 1 R2. Densitometric analysis is shown as means  $\pm$  SD, n = 4 mice per group (2 in Figure 1 R2 and 2 in this figure). \*\*\*P < 0.001, \*\*P < 0.005, \*P < 0.05 wild-type vs Zmpste24<sup>-/-</sup> mice.



Appendix Figure S3. Densitometry of Figure 2 R2. Densitometric analysis is shown as means  $\pm$  SD. Data represent 3 different experiments from the same patient. \*\*\* $P < 0.001$ , \*\* $P < 0.005$ , \* $P < 0.05$  control cells vs patient cells; <sup>aaa</sup> $P < 0.001$ ; <sup>aa</sup> $P < 0.01$ ; <sup>a</sup> $P < 0.01$  no treatment vs treatment; <sup>bb</sup> $P < 0.005$ ; <sup>b</sup> $P < 0.05$  THP cells no treatment vs treatment.



Appendix Figure S4. Effect of the MCC950 treatment in skin fibroblasts from HGPS patient 2. Data represent 3 different experiments from the same patient. \*\*\* $P < 0.001$ , \*\* $P < 0.005$ , \* $P < 0.05$  control cells vs patient cells; <sup>aaa</sup> $P < 0.001$ ; <sup>aa</sup> $P < 0.01$ ; <sup>a</sup> $P < 0.01$  no treatment vs treatment.