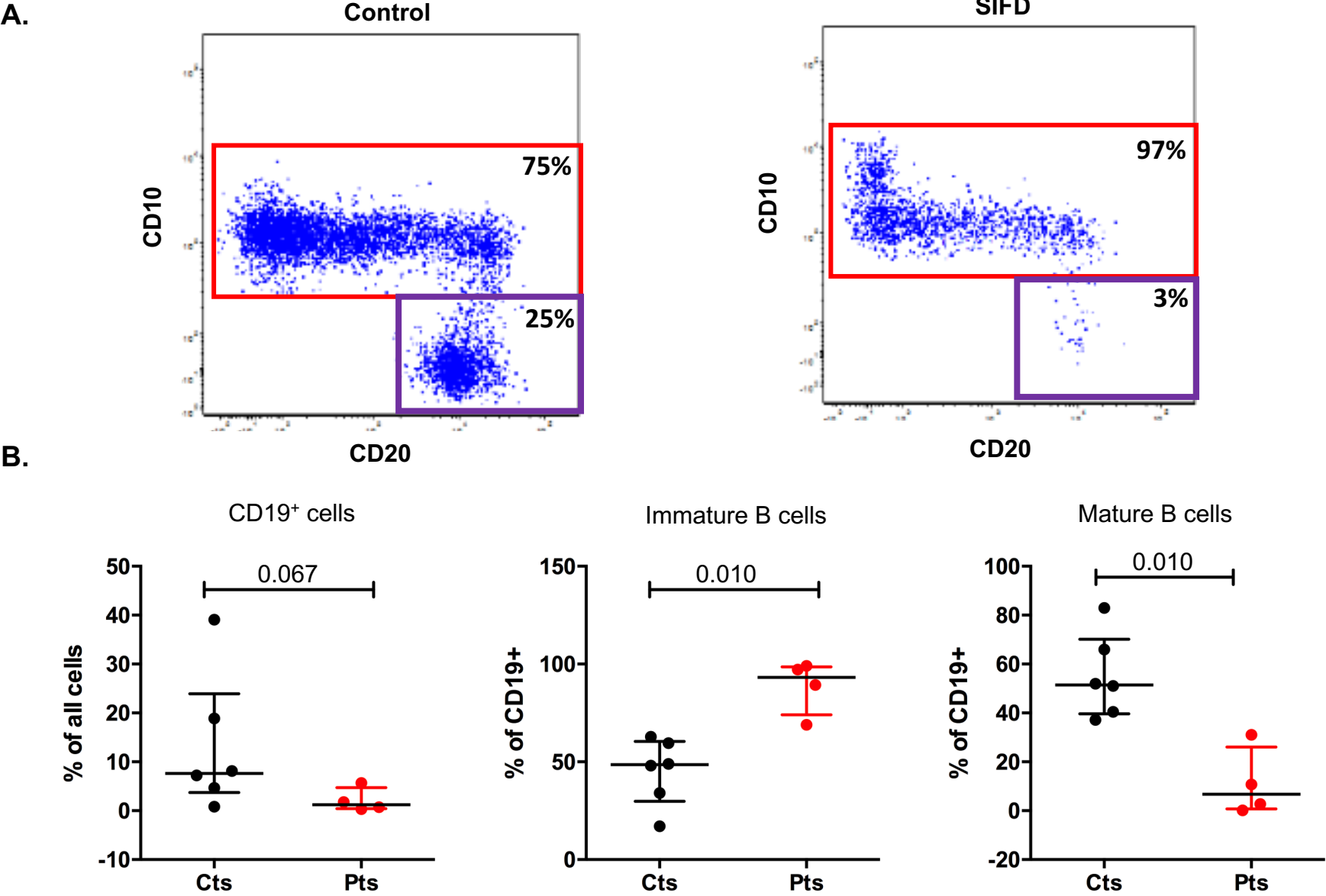


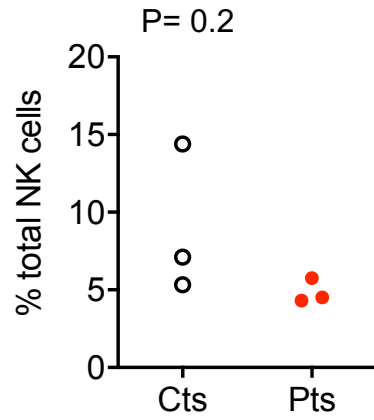
Supplementary figure 1. Impaired B-cell maturation in the bone marrow of SIFD patients



Cts n=6; Pts n=4

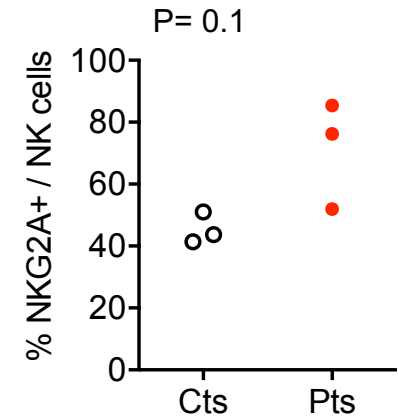
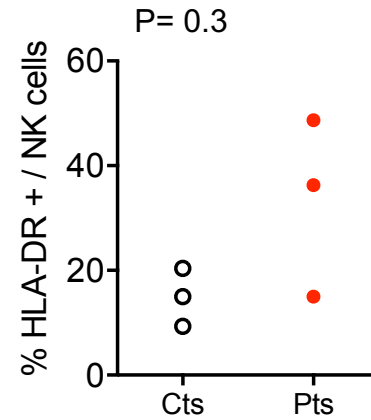
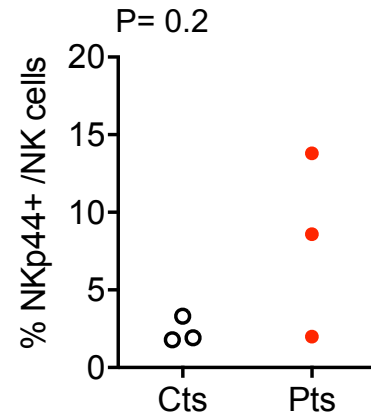
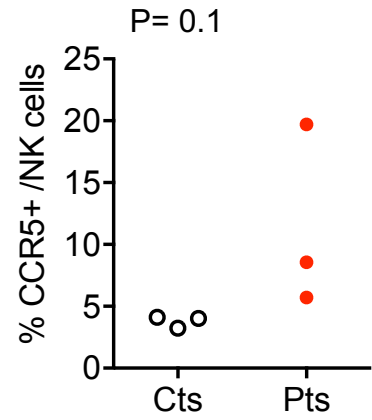
Supplementary figure 2. NK cell phenotype and function

A.

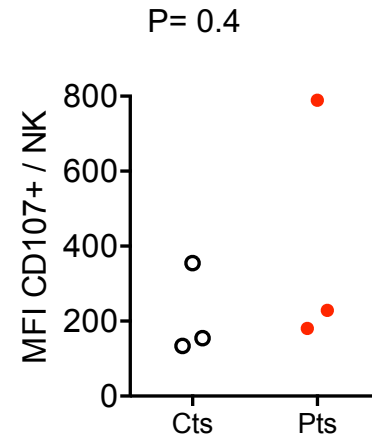
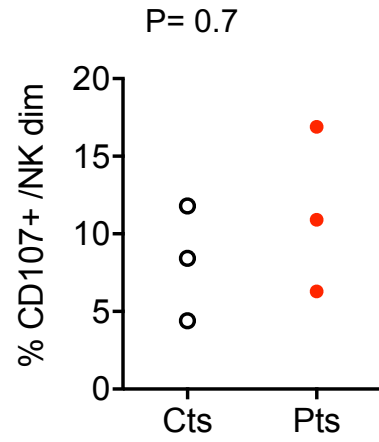
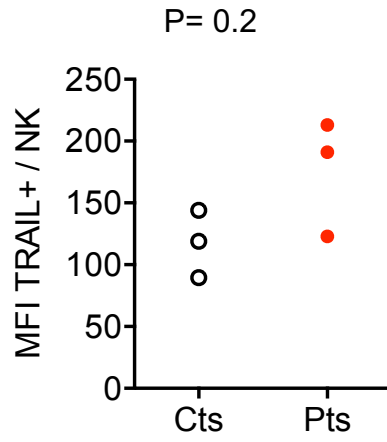
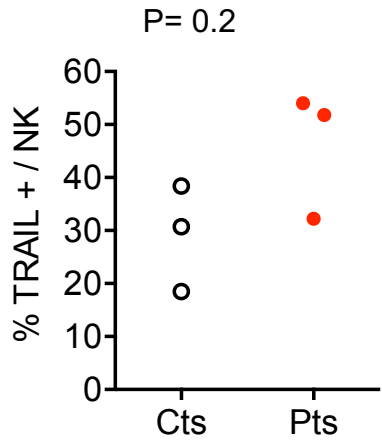


Cts n=3 Pts n=3

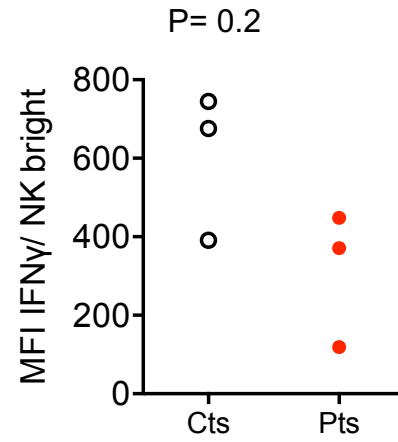
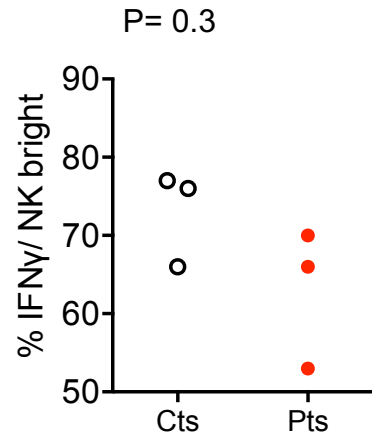
B.



C.



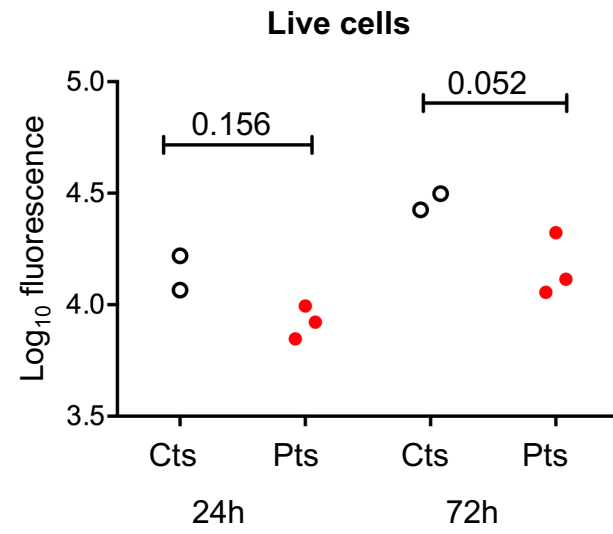
D.



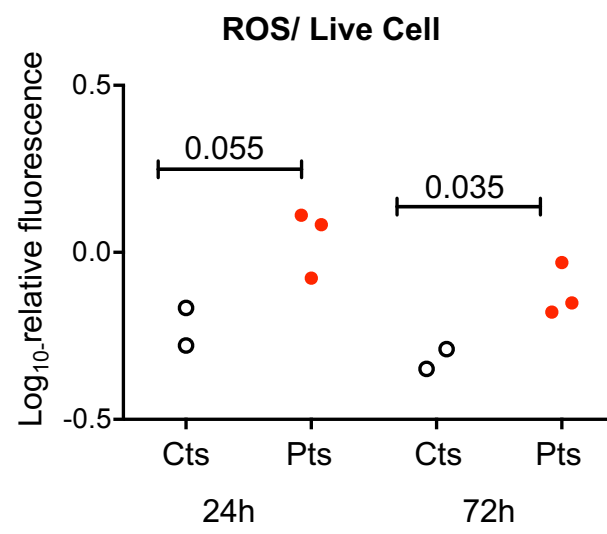
Cts n=3 Pts n=3

Supplementary figure 3. ROS overproduction

A.

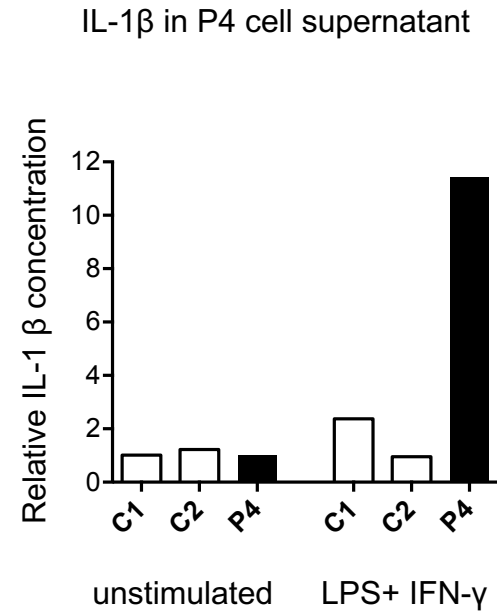


B.

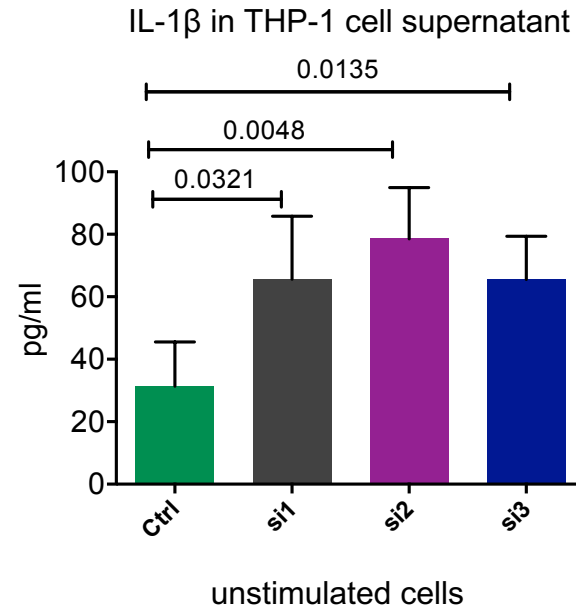


Supplementary figure 4. IL-1 β production

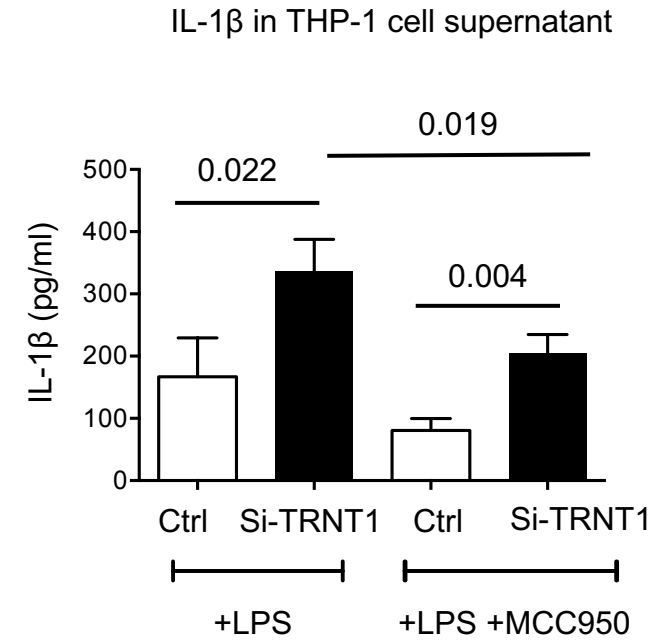
A.



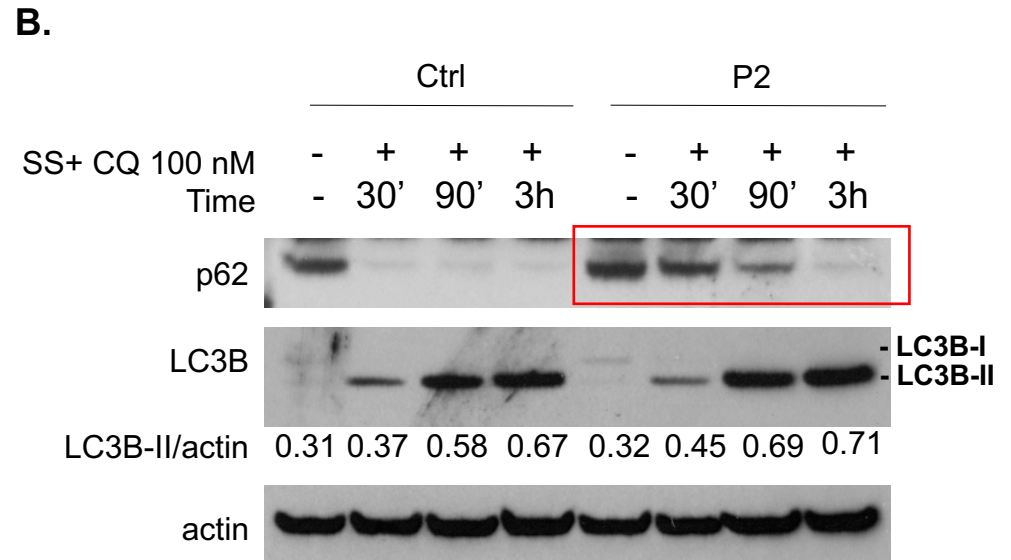
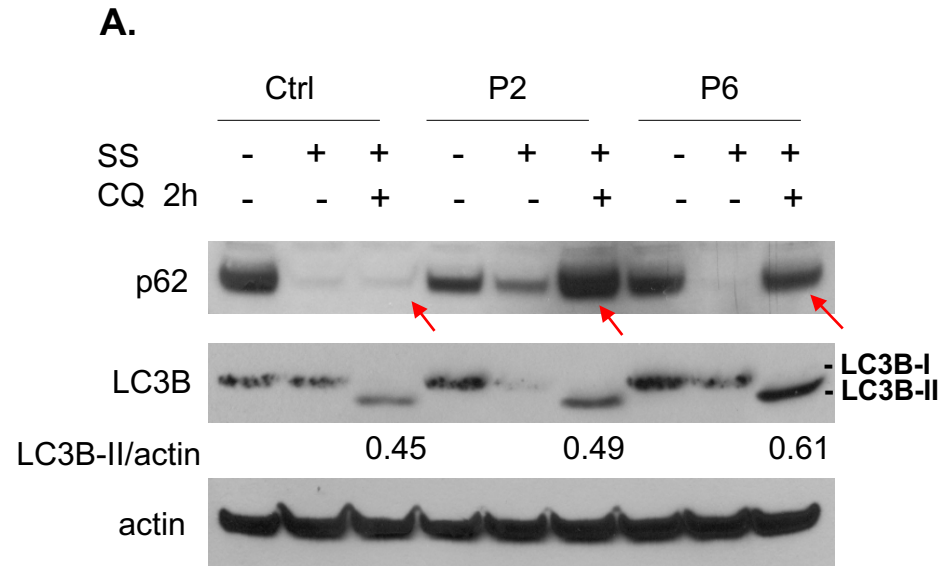
B.



C.

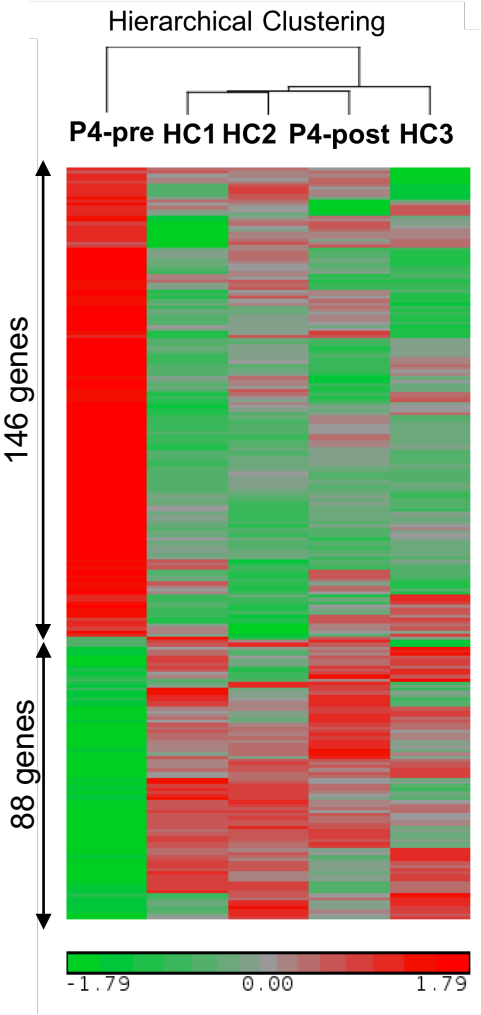


Supplementary figure 5. Defective degradation of the ubiquitin-scaffold protein, p62, in mutant cells

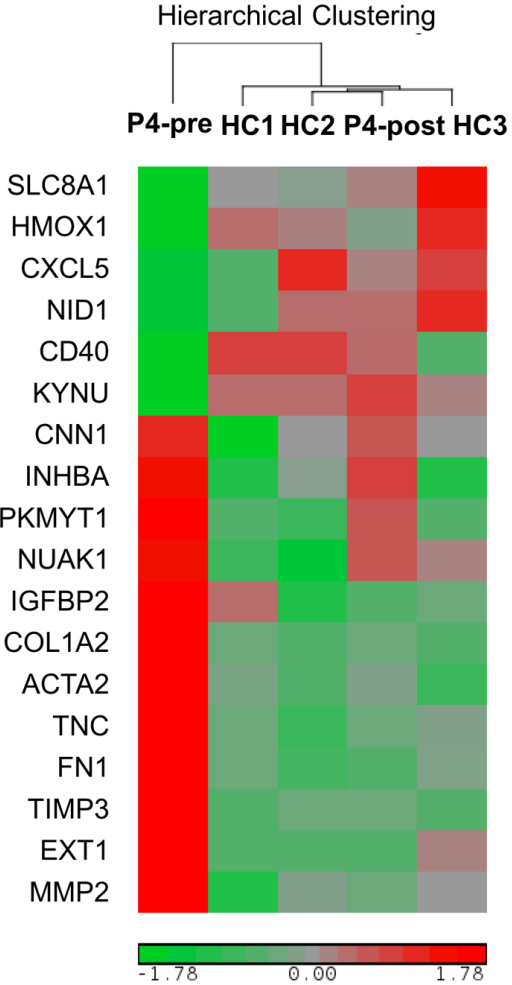


Supplementary figure 6. Changes in gene expression after treatment with TNF-inhibitor

A.

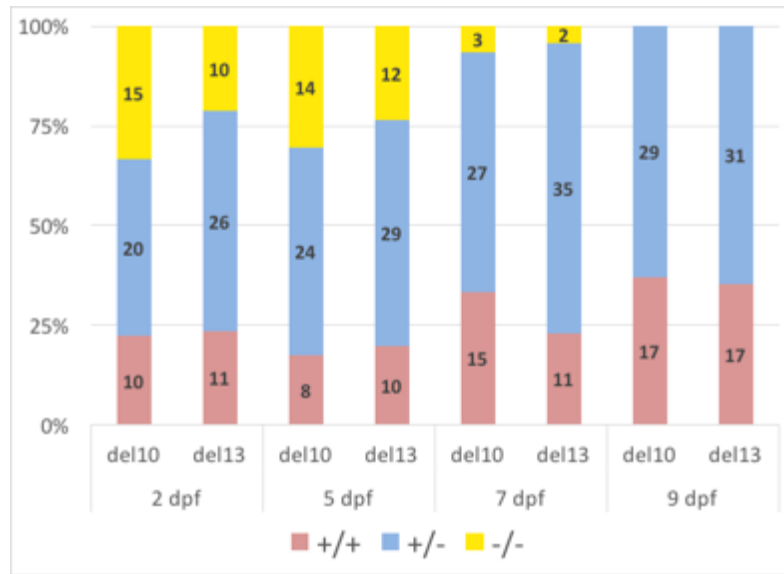


B.

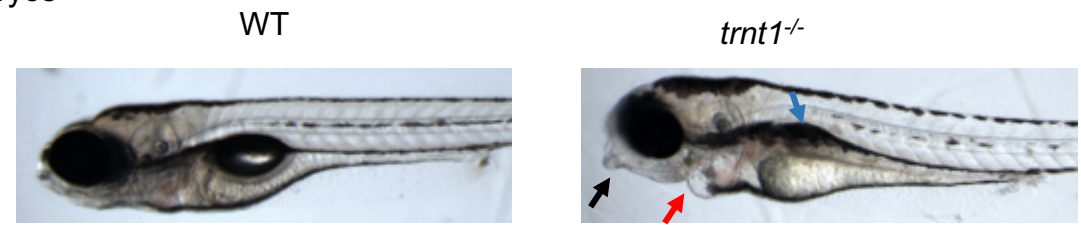


Supplementary figure 7. Viability and phenotype of *trnt1* knockout zebrafish embryos

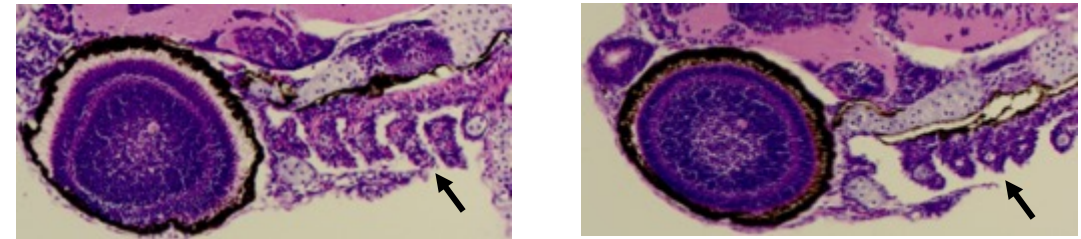
A.



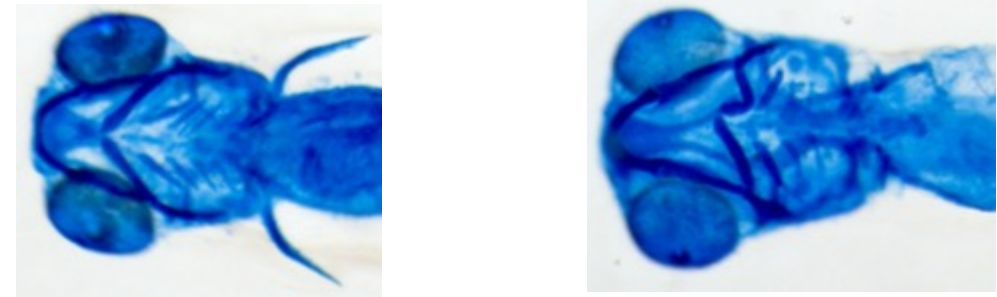
B.



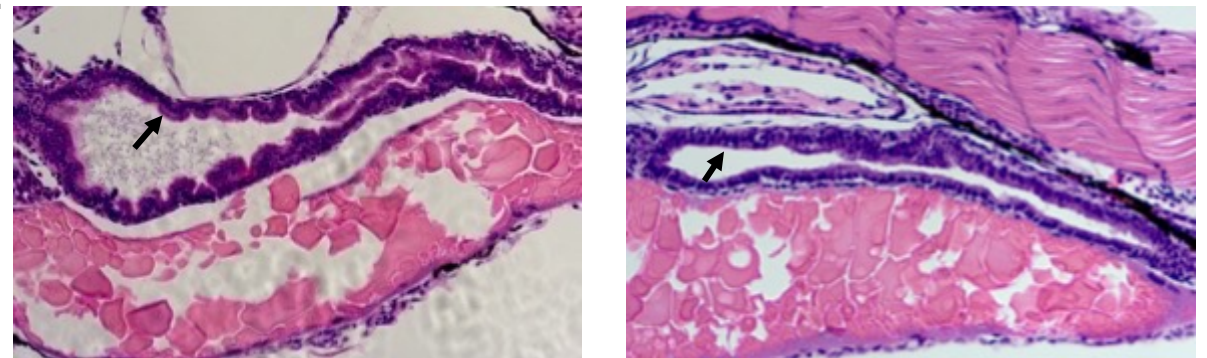
C.



D.



E.



Supplementary figure 8 . Reduced neuromast hair cells in *trnt1* mutants

