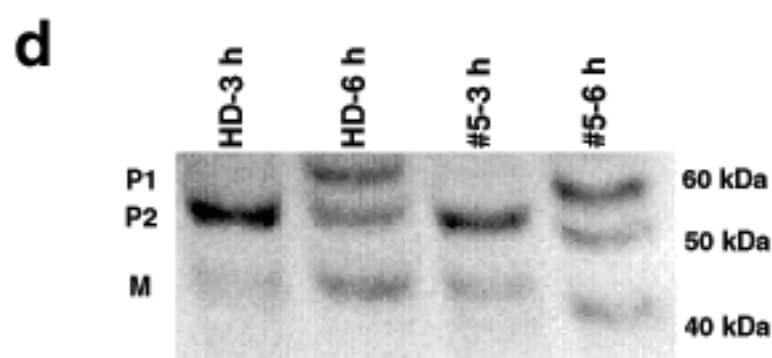
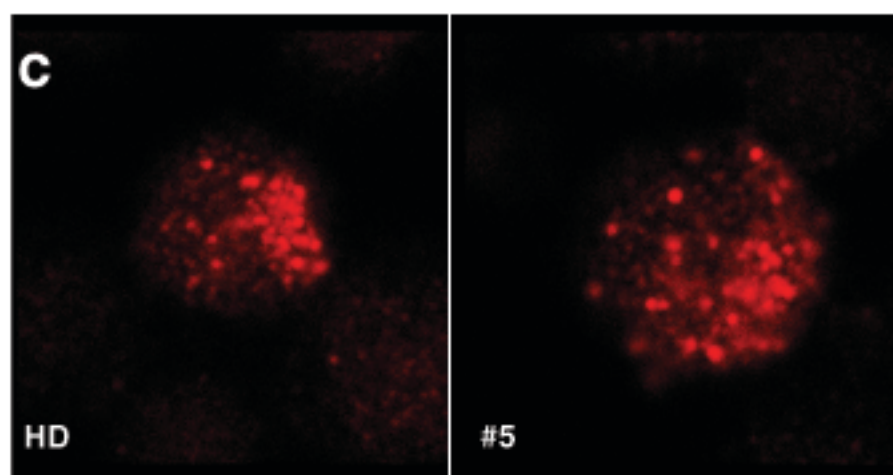
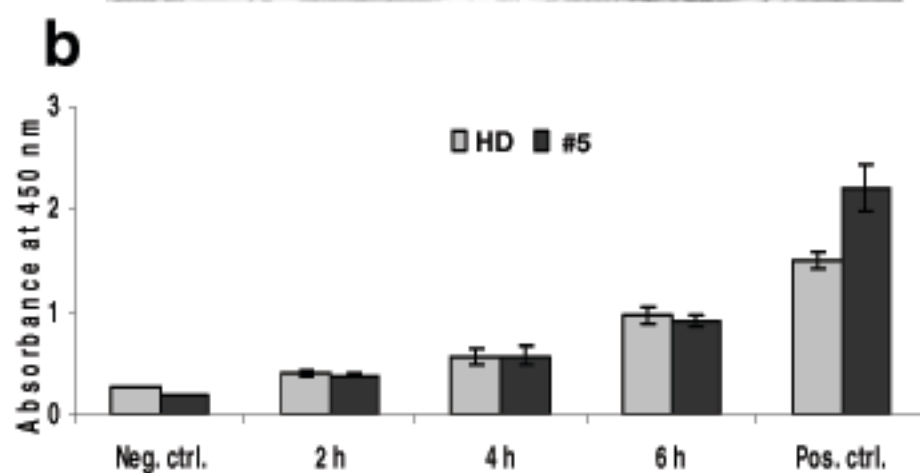
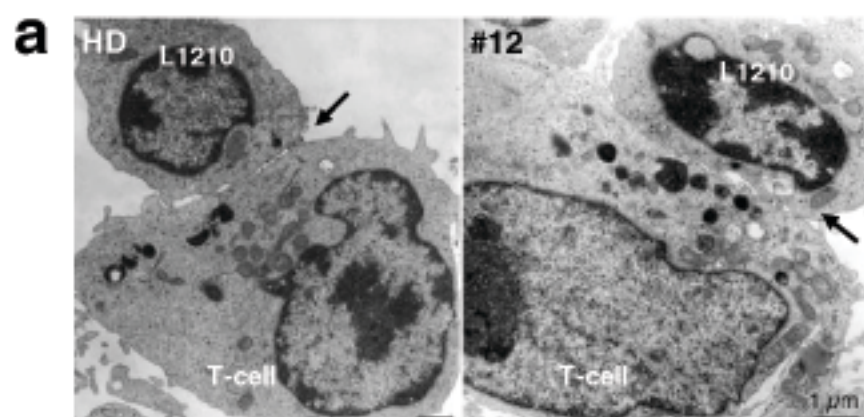
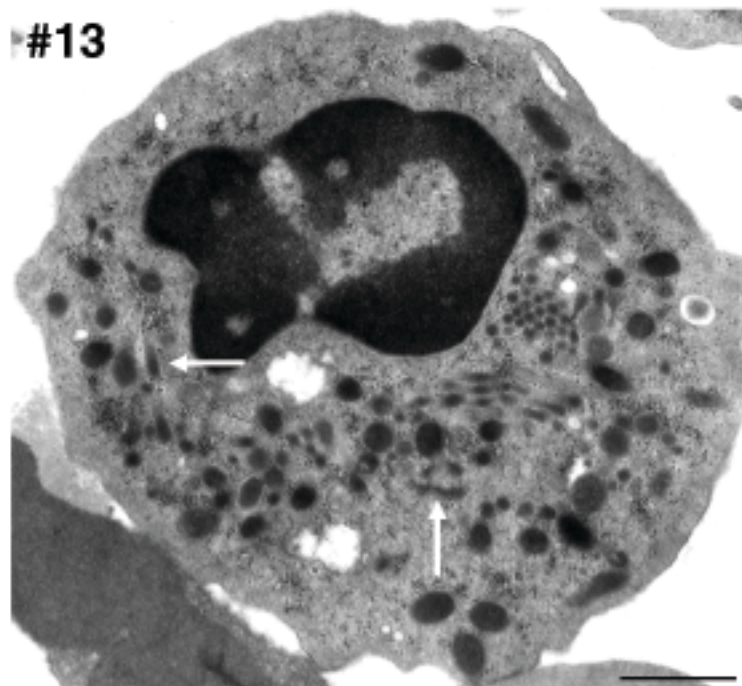
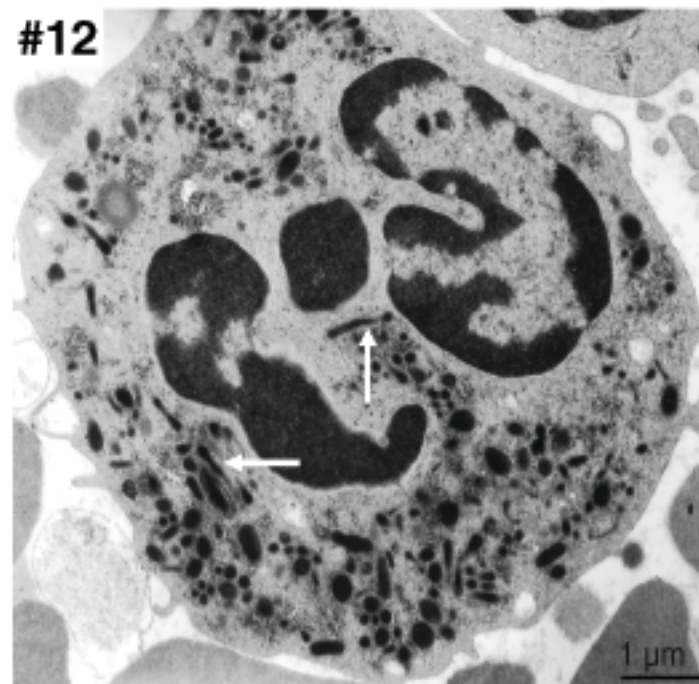
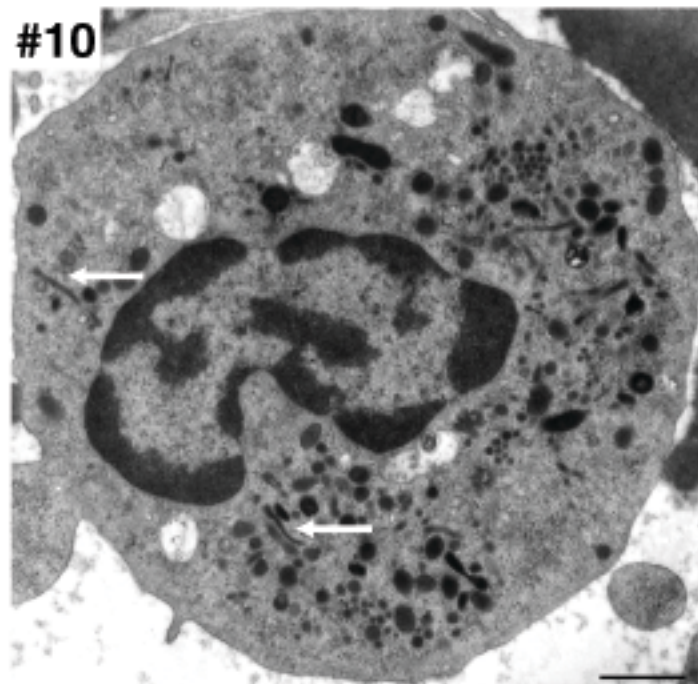




e



Supplementary Figure 1 Assessment of granzyme B and perforin in cytotoxic T-cells, and TEM of neutrophil granulocytes of patients #10, 12 and 13. **(a)** TEM of T-cell and L1210 target cell interaction. Secretory granules localize to the immunological synapse (arrow) both in patient (#12) and control T-cells (HD). **(b)** ELISA assay measuring granzyme B release in supernatant of OKT3-stimulated CD8⁺ T-cells. Maximum release was measured upon TritonX-induced cell lysis. **(c)** Intracytoplasmic staining of perforin in CD8⁺ T-cells. **(d)** Western blot analysis of perforin in CD8⁺ T-cells from patient (#5) and healthy donor (HD). Cells were stimulated on OKT-3 for 3 and 6 h as indicated. P1 and P2 correspond to two processed forms of perforin, M denotes mature perforin. For details see **Supplementary Methods** online. **(e)** TEM of neutrophil granulocytes of patients #10, 12 and 13. Abnormally shaped, elongated azurophilic granules are indicated by arrows. scale bar: 1 μ m.