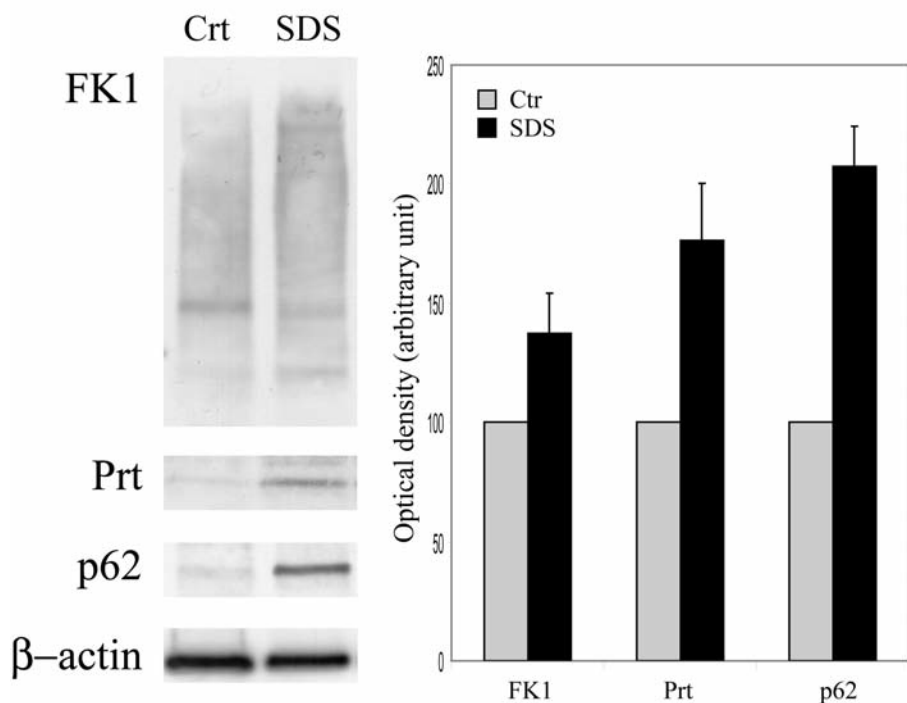


Ubiquitin-proteasome-rich cytoplasmic structures in neutrophils of patients with Shwachman-Diamond syndrome

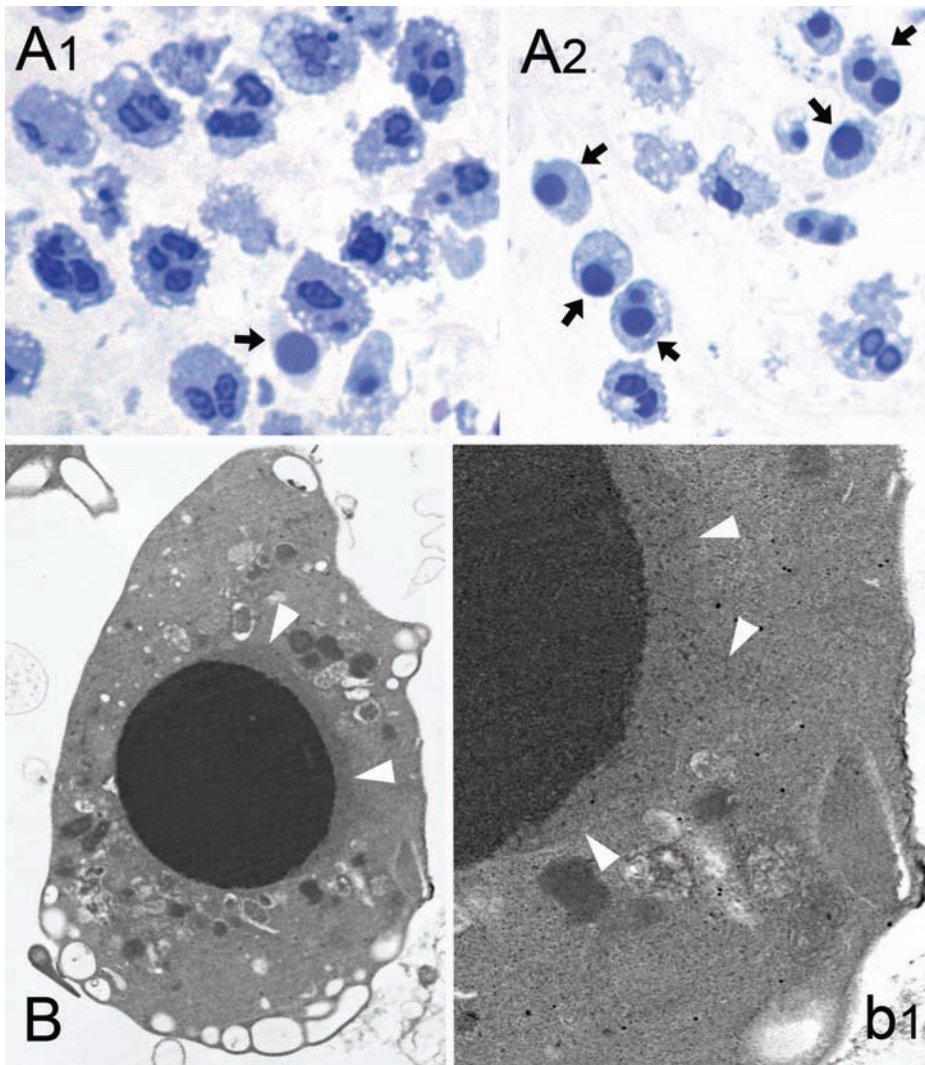
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Online Supplementary Figure S1. Representative western blot of lysed blood granulocytes from a healthy control subject and an SDS patient with *SBDS* gene mutation. Lysates were resolved by SDS-PAGE (4–20%) and immunoblotted for FK1 (polyubiquitinated proteins), 20S proteasome (Prt) and p62 protein. β-Actin was used as a loading control. The graph on the right, obtained by densitometry and normalized to β-actin, shows the increment, expressed as a percentage, of FK1, Prt and p62 in SDS patients compared to that in control granulocytes.



Online Supplementary Figure S2. Aldehyde-osmium-fixed blood neutrophils from a healthy control, either untreated (**A1**, 1,000x) or treated with the xanthine/xanthine oxidase system (**A2**, 1,000x; **B**, 4,000x; **b1**, 20,000x). In toluidine-blue-stained semithin resin sections a single cell (black arrow) in **A1**, and several cells in **A2**, show round, pyknotic nuclei. The apoptotic nature of these cells was confirmed by electron microscopy of an adjacent thin section (**B**, enlarged in **b1**) showing a round dense, chromatin aggregate, leaving a thin crescentic remnant of karyoplasm in direct contact with a mostly homogeneous cytoplasm, at a thin borderline (white arrowheads) devoid of a nuclear membrane envelop. A few secretory granules, dense bodies and several membrane-delimited vesicles, but no PaCS, can still be recognized in the cytoplasm.