Supplementary material to article by M. El Hachem et al. "Structural Defects of Laminin  $\beta$ 3 N-terminus Underlie Junctional Epidermolysis Bullosa with Altered Granulation Tissue Response"



*Fig. S1.* Molecular findings in junctional epidermolysis bullosa (JEB) with altered wound repair. (a) DNA sequence analysis of the region spanning *LAMB3* codon 254 (*underlined*). The G-to-A nucleotide transition (c.761G>A) is indicated by an *arrow.* (b) Schematic of laminin-332 heterotrimer and modelling of superimposed wild-type and mutant structures of the  $\beta$ 3 LN-LE1-4 domains with mutation p.Gly254Asp, shown in *red spheres.* LN domain in *magenta*, LE1 domain in *green.* (c) Ramachandran map of aspartates. For this analysis, high-quality protein structures were downloaded from the Protein Data Bank (PDB), and the phi and psi angles were computed for all aspartate residues in those selected structures. The distribution of the phi and psi angles of the aspartate residues is shown as a contour plot, in which the colour gradient represents the frequency of every angle's combination (darker yellow intensity indicates a more favoured conformation). Asp254 in the mutant model of the laminin structure, shown as a *red dot (arrow)* in the graph, is in a very low-density zone of the Ramachandran map; thus in a very unfavourable phi/psi conformation. Indeed, more than 99% of the aspartate residues of PDB structures are in a more favoured phi/psi conformation.