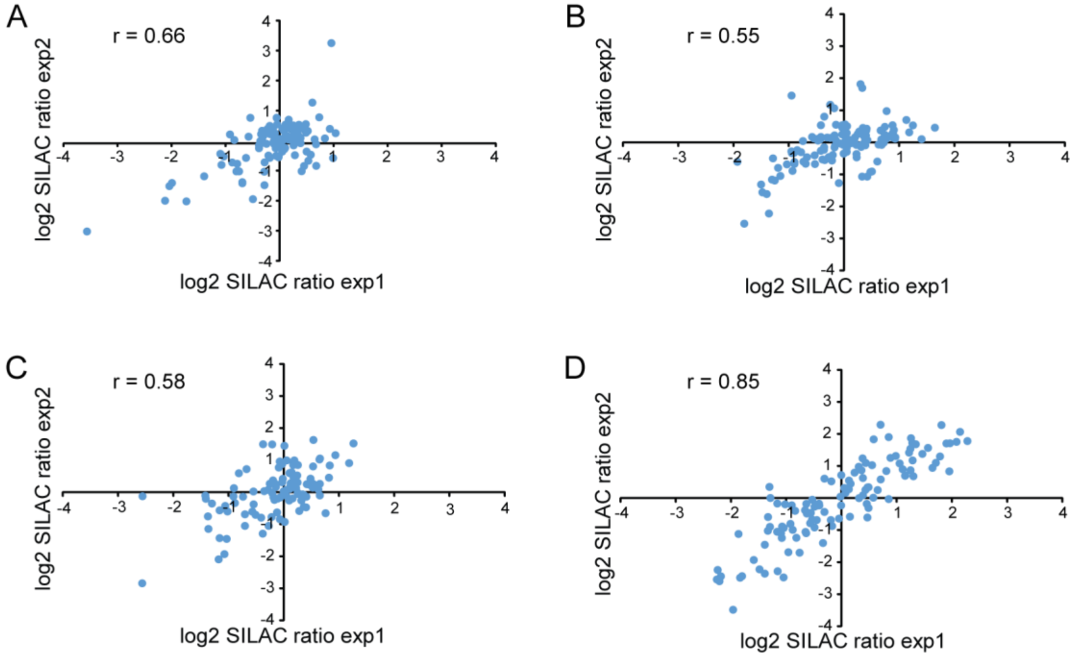
Combinatorial omics analysis reveals perturbed lysosomal homeostasis in collagen VII-deficient keratinocytes

Kerstin Thriene, Björn Andreas Grüning, Olivier Bornert, Anika Erxleben, Juna Leppert, Ioannis Athanasiou, Ekkehard Weber, Dimitra Kiritsi, Alexander Nyström, Thomas Reinheckel, Rolf Backofen, Cristina Has, Leena Bruckner-Tuderman, Jörn Dengjel

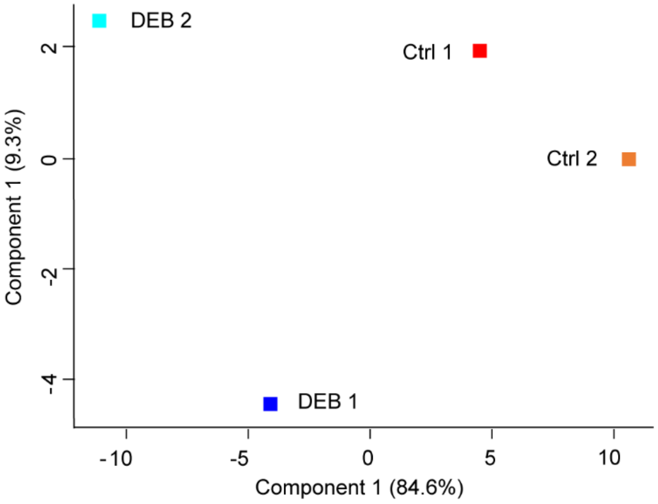
Supplemental Figures

Supplemental Figure S1: Correlation of replicates of ECM isolated from primary human keratinocytes.



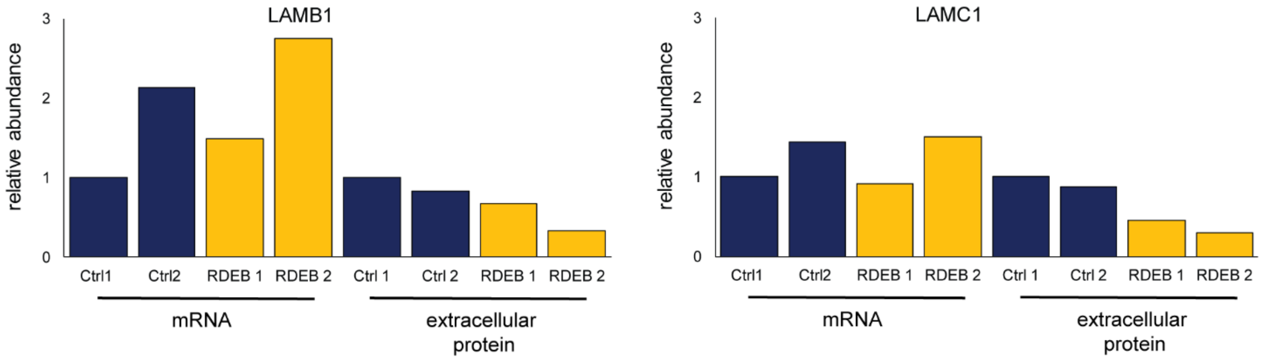
Correlation of biological replicates of the ECM proteome isolated of primary human DEB keratinocytes (A, B) or healthy controls (C, D), r = Pearson correlation coefficient.

Supplemental Figure S2: PCA replicates ECM



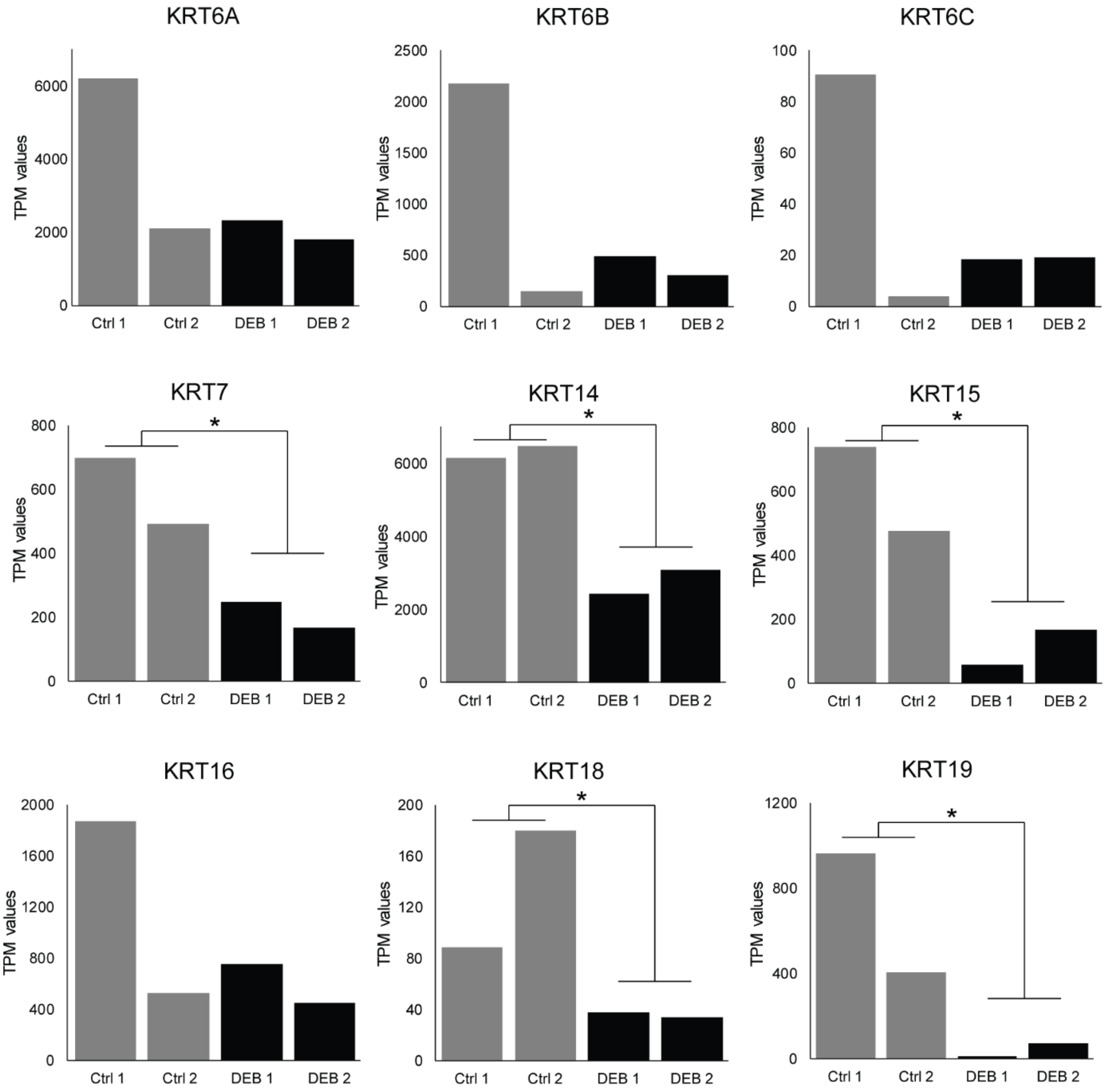
Principle component analysis (PCA) of combined SILAC ratios for each patient and the respective controls.

Supplemental Figure S3: Relative mRNA and protein abundances of laminin beta 1 and gamma 1



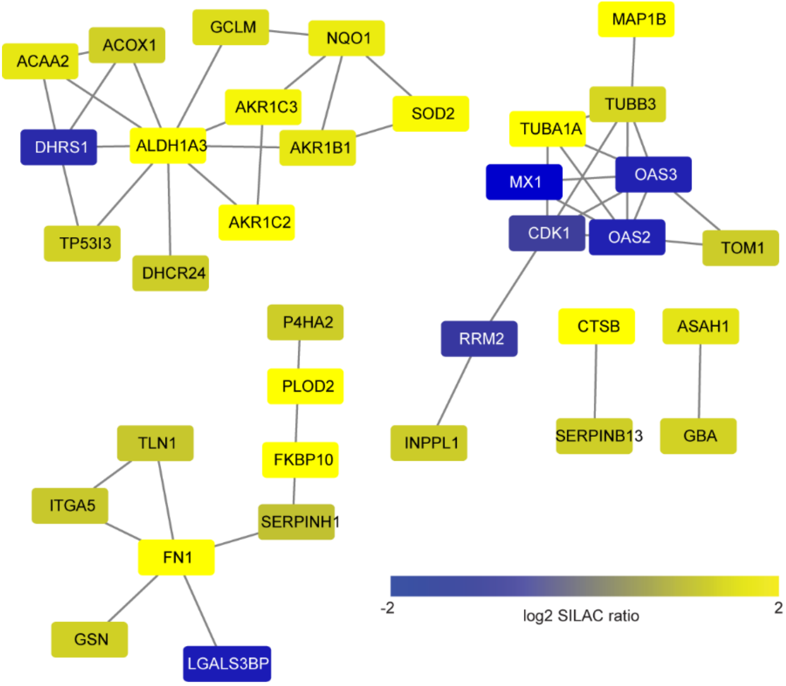
Relative values compared to the respective control 1 (Ctrl 1) are shown. Protein levels were significantly reduced (Welch's t-test, Permutation-based FDR 0.05) in ECM isolated of DEB cells compared to controls while mRNA levels did not change significantly.

Supplemental Figure S4: Gene expression analysis of keratin-encoding genes.



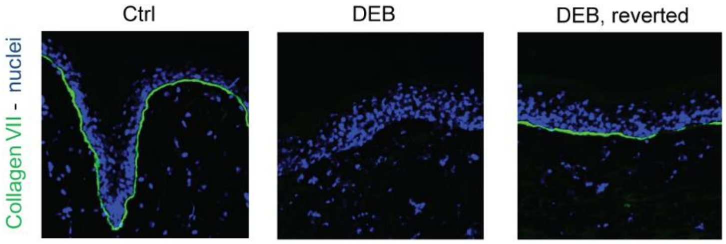
Gene expression analysis of keratin encoding genes. Bar graphs show transcript abundances (TPM values). *: BH-corrected q-value < 0.1.

Supplemental Figure S5: Network analysis of significantly regulated intracellular proteins.



STRING DB was used to identify protein-protein interactions of significantly regulated proteins (confidence score 0.4; Welch's t-test, Permutation-based FDR 0.05, minimal fold change > +/- 2). Color scale indicates log2 transformed SILAC ratios of respective proteins.

Supplemental Figure S6: Spontaneous restoration of *COL7A1* expression in skin of a DEB patient



Immunofluorescence staining of DEB skin reveals spontaneously restored C7 levels (green signal). Blue = DAPI staining.