2.06 Induced pluripotent stem cell (iPSC)-derived fibroblasts for the study of Systemic Sclerosis (SSc)

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Background: Systemic Sclerosis (SSc) is a rare autoimmune disease, mainly affecting the skin. 40% of SSc patients develop Interstitial Lung Disease (SSc-ILD), a disease with higher mortality risk. The mechanisms underlaying this transition are unknown. **Objective:** Using peripheral blood Induced Pluripotent Stem Cells (iPSCs) differentiated into fibroblasts, we aim to identify differences between SSc and SSc-ILD fibroblasts. **Methods**: Peripheral blood-derived mononuclear cells from one patient with SSc, one with SSc-ILD and a healthy control were reprogrammed into iPSCs and differentiated into fibroblasts. Total RNA was collected on day 3/6 and at subsequent passages. Cells were characterized using qRT-PCR. **Results**: The iPSCs were successfully transformed into fibroblasts. The two SSc/SSc-ILD cell lines showed significant fibroblasts markers (Col1A1, Vimentin, S100A4) from differentiation day 6. At p4 both cells showed similar levels of Vimentin and S100A4 expression, but differences in COL1A1 and CXCL12 mRNA expression. Sirius Red staining confirmed the difference in collagen. **Conclusions**: We successfully transformed blood-derived mononuclear cells from patients with SSc and SSc-ILD into fibroblasts. SSc and SSc-ILD fibroblasts may have some key behavioural and physiological differences which could help explain the ***progression to SSc-ILD.*** Further work is required to characterise the cells' TGFβ_1 response and disease specific behaviour. **Conflicts of interest:** Authors declare no conflicts of interest