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J Tissue Sci Eng 2017, 8:3 (Suppl)
DOI: 10.4172/2157-7552-C1-037

Disease modeling of systemic sclerosis disease through iPS induction and differentiation

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Systemic sclerosis is a connective tissue disease, characterized by immunological abnormalities, vascular injury and fibrosis of the skin and various internal organs. The pathogenesis of scleroderma is unknown, but it seems apoptosis and vascular alterations can be considered as the first signs of systemic sclerosis appearance. Therefore finding out the endothelial alteration mechanism is one aspect of disease progress study. These alterations can be investigated in 2 steps to check if these endothelial cells have any difficulties in their differentiation or lacks necessary factors for angiogenesis. To answer these questions, we need a good model to mimic these cells development and differentiation in vitro. Isolation of these cells from patients are not always possible or enough, but substantially stem cell therapy represents a hope for differentiation study based on the ability of pluripotent stem cells to differentiate into all three germ layers including endothelial cells in vitro. In comparison with matured, transformed, primary or engineered cells, pluripotent embryonic stem cells (ESCs) have unique characteristics and advantages in development of important cell-based tools. However, availability of human ESCs from patients with specific medical needs is a major limitation of these cells. Induced pluripotent stem (iPS) cells, which are derived from somatic cells, may represent a potential strategy to overcome the limitations ESCs.

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