**Background**

Idiopathic pulmonary fibrosis (IPF) is a chronic disease and the most common form of idiopathic interstitial pneumonia. IPF is a heterogeneous disease with areas of subpleural and paraseptal fibrosis and honeycombing (clustered cystic airspaces with delicate walls). It is often fatal, and the time from diagnosis to death is variable. The hallmark of IPF is the presence of fibroblast foci, which are regions of fibrosis surrounded by a rim of fibroblasts. These foci are characterized by increased collagen deposition and decreased thickness of airspaces.

**Introduction**

Several patients with peroxisomal biogenesis disorders develop fibrosis of the lung during their first year of life and are not studied in this study. One of the patients presented by Talmadge et al. (2011) had idiopathic pulmonary fibrosis. PEX13 is an essential gene for peroxisome biogenesis and is involved in the pathogenesis of IPF.

**Methods**

Collagen deposition and other extracellular matrix (ECM) proteins characteristic of the current ongoing disease. Collagen deposition and other extracellular matrix proteins are increased in fibroblasts treated with interferin control. The expression of 28SrRNA and the HPRT1 genes was used as controls. Catalase was targeted to cytoplasm in PEX13 KO, revealing a targeting defect whereas only small differences were observed in wild-type and PEX13 siRNA-treated fibroblasts.

**Results**

The number of cells per square in each different sample is depicted.

**Discussion**

The strong down-regulation in peroxisomal eicosanoid degradation might lead to the accumulation of proinflammatory mediators and the peroxisomal compartment was not dependent on peroxisomal metabolic proteins might be dysregulated in IPF and being one of the reasons leading to the aggravation of the pulmonary disease. The expression of PEX13 and the HPRT1 genes was used as controls. Catalase was targeted to cytoplasm in PEX13 KO, revealing a targeting defect whereas only small differences were observed in wild-type and PEX13 siRNA-treated fibroblasts.

**Conclusion**

In conclusion, catalase the peroxisomal enzyme with strongest capacity to degrade hydrogen peroxide was only mildly affected in fibroblasts of IPF patients, even though the effect on antioxidant enzyme activities was observed. The expression of 28SrRNA and the HPRT1 genes was used as controls. Catalase was targeted to cytoplasm in PEX13 KO, revealing a targeting defect whereas only small differences were observed in wild-type and PEX13 siRNA-treated fibroblasts.