特發性肺纖維化最新臨床證據

An update of IPF

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Idiopathic pulmonary fibrosis (IPF) is a progressive, irreversible, and typically fatal lung disease characterized by subpleural fibrosis, subepithelial fibroblast foci, and microscopic honeycombing. There has been a recent influx of new observations aimed at explaining the mechanisms responsible for the initiation and progression of pulmonary fibrosis. However, despite this, the pathogenesis of the disease is largely unclear. Recent progress has been made in the characterization of specific pathologic and clinical features that have enhanced the understanding of pathologically activated molecular pathways during the onset and progression of IPF.

IPF has an unknown etiology and matures rapidly, while being characterized by acute lung injury with consequent scarring on the pleura. The future demands a better clinical evaluation of IPF, and the indispensable developments in coming years will likely see a widely explored inclusive diagnostic criterion. The diagnostic procedures have seen use of non-standardized clinical information including multidisciplinary team discussion based on individual case to case study. A better diagnosis approach will oversee the permutations and combinations of non-standardized data combined with new rigid diagnostic criteria. Thus, the most likely future multidisciplinary model will be comprised of a conjunction of better diagnostic applications augmented by nonstandardized clinical reasoning based on individual patient information.

Treatment at present is focused on the interruption of the diseases progress by targeting specific cellular mechanisms that are tied to IPF. It is imperative to have a better understanding of the cellular mechanism of IPF for the development of future drug therapies. As such, future directives should encourage the search for new molecular targets for drug therapy, refinement of already available treatments for a better efficacy with less side effects and research into the identification of genetic susceptibility factors. In view of this, future effective therapy regimens therapy will rely heavily on selection of drug agents that fits to individual genetic makeup, biomarker profiles and responses to environmental triggers. This speech will be highlights several of the advances that have been made and focus on the pathobiology of IPF. Finally, a complete repertoire of the treatment therapies that have been used in the past as well as future medications and therapies is provided.