inhale air: the lung loses its compliance and the airway resistance increases (compliance = 1/resistance). As the disease progresses there is also an increase in vascular resistance. The site of action of Treprostinil in pulmonary fibrosis is the vasculature and the interstitial space in the alveola.

Tissieres et al. describe studies using iloprost for the treatment of a patient with cystic fibrosis and secondary pulmonary hypertension. It is disclosed that aerolised iloprost was effective in lowering pulmonary artery pressure (The annals of thoracic surgery, vol, 78, no.3, E48-E50).

US2001/006979 A1 describes the use of prostacyclin derivatives like iloprost or cicaprost for the treatment of fibrotic diseases.

Cystic fibrosis is unrelated to pulmonary fibrosis because it is a disease that originates in the bronchial epithelium. Because of the absence of CFTR, there is too little water in the mucus that covers the bronchial epithelium; accordingly, the cilia cannot move the thick mucus and mucociliary clearance breaks down (mucociliary clearance works like a conveyor belt, where the cilia beat rhythmically in a concentred manner to move the mucus back to the trachea and pharynx, from where it may be cleared by swallowing or coughing etc.). If mucociliary breaks down, the bacteria cannot be removed from the bronchi, the bronchi are colonized by bacteria and there are repeated bouts of lung infections that destroy the lung. The situation can be remedied by restoring Cl- fluxes to the bronchial epithelium. Thus, in cystic fibrosis the site of action is the airway epithelium of the bronchi. The site of action is anatomically distinct (lung interstitium vs. bronchial airway), involves a different set of cells (fibroblasts, vascular smooth muscle cells, endothelium versus absorbing and secreting bronchial epithelial cells) and presumably also involves different receptors (prostacyclin receptor vs possibly EP2-receptor).

Presently, no treatments of cystic fibrosis are available that significantly improve quality of life of patients over a longer period. Therefore it is an object of the present embodiments disclosed herein~~invention~~ to provide compositions for treatment that can enhance the expression of ΔF508 CFTR and/or chloride channel function in epithelial cells of the lung.

Short description ~~of the invention~~:

The object of the present embodiments disclosed herein~~invention~~ is achieved by providing a composition comprising a prostacyclin or an analogue, a derivative or a pharmaceutically acceptable salt thereof for use in preventing or treating cystic fibrosis.

In one aspect, there is provided the use of a composition comprising a prostacyclin or analogue or derivative or a pharmaceutically acceptable salt thereof in the manufacture of a medicament for preventing or treating cystic fibrosis wherein said prostacyclin analogue is selected from the group consisting of Treprostinil, Iloprost, Cisaprost, Beraprost, derivatives and pharmaceutically acceptable salts thereof.

According to a specific embodiment of the disclosure~~invention~~ the prostacyclin analogue is selected from the group of Treprostinil, Iloprost, Cisaprost or Beraprost or pharmaceutically acceptable salts thereof.

More specifically, the Treprostinil derivative can be selected from the group of acid derivatives, prodrugs, sustained release forms, inhaled forms, oral forms, polymorphs or isomers of Treprostinil.

The composition of the disclosure~~invention~~ can be administered by intravenous administration, inhalation or it can be in an orally available form selected from the group of tablets or capsules.

Specifically the composition comprises an effective amount of Treprostinil or its derivative, or a pharmaceutically acceptable salt thereof which is at least 1.0 ng/kg of body weight/min.

Further, according to the present disclosure~~invention~~ the use of a kit for treating or preventing a condition associated with cystic fibrosis in a subject, comprising (i) an effective amount of a prostacyclin or prostacyclin analogue or derivative or a pharmaceutically acceptable salt thereof, specifically a pharmaceutically acceptable salt of Treprostinil, (ii) one or more pharmaceutically acceptable carriers and/or additives, and (iii) instructions for use in treating or preventing cystic fibrosis is provided as well.

Figures:

Fig. 1: Accumulation of cAMP in IB3-1 cells after incubation with Treprostinil

Fig. 2: Activation of a Cl-current by Treprostinil in the human bronchial epithelial IB3-1 cell line transiently expressing CFTR-wt.