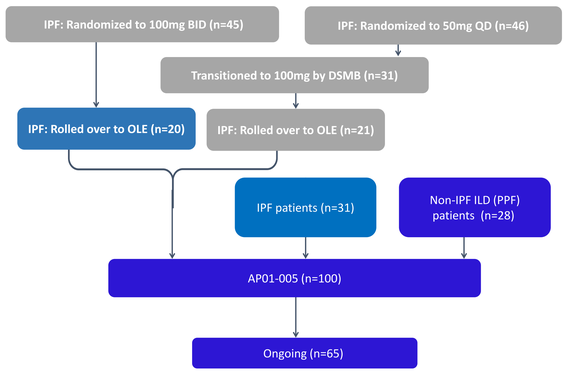
Nebulised pirfenidone in non-idiopathic pulmonary fibrosis (IPF) progressive pulmonary fibrosis (PPF): first look at FVC data

Dr A West[1], Dr A Lawrence[1], H Bao[2], D Nair[2], C Thompson[2], Dr F Woodhead[2]
[1]Guy's & St Thomas' NHS Foundation Trust- London (UK), [2]Avalyn Pharma Inc.- Seattle, Washington (USA)

Background

Oral antifibrotics attenuate the decline of lung function in patients with **progressive pulmonary fibrosis (PPF)** and oral Nintedanib is now considered standard of care. Side-effects, particularly gastrointestinal, are often reported with Nintedanib, and may lead to dose reduction or limitation of treatment.

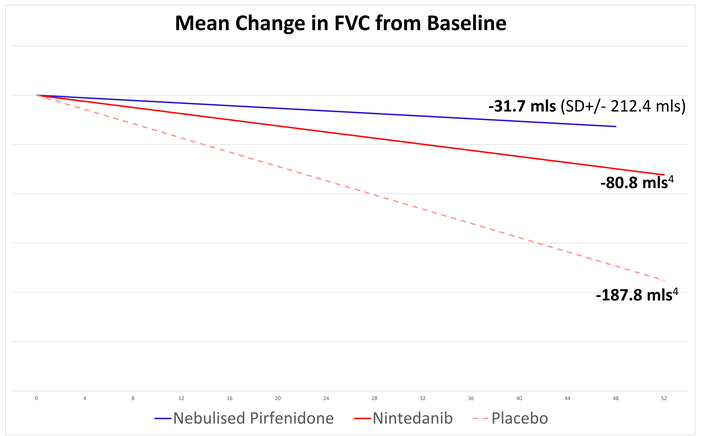
Nebulised Pirfenidone has been shown to be safe[1]. It has also been shown to achieve both **approximately x35 peak epithelial lining fluid concentration (C_{max})** with **<1/15th systemic absorption**[2,3] of standard dose oral pirfenidone. This suggests the nebulised route has the potential both for effectiveness and improved tolerability.



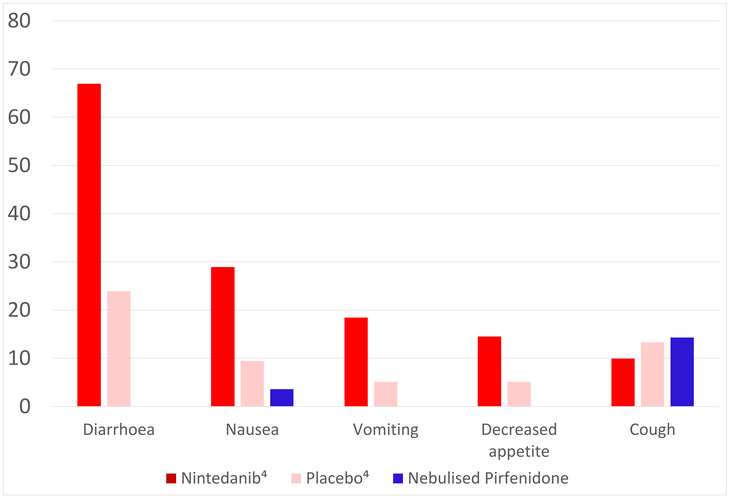
The **ATLAS** open-label extension was open to patients with **PPF**, and was designed to assess the safety of nebulised Pirfenidone. Patients were included if they had **chronic progressive fibrotic ILD** without treatment alternatives.

Baseline Characteristics & FVC Data

Baseline Characteristics	(n=28)
Age	63.8 yrs (SD+/-11.59)
Male : Female	12: 31
Baseline FVC	
<65%	39%
>65 to <80%	29%
>80%	32%
Mean FVC	75.3%p
Diagnoses	
CHP	14.3%
CTD-ILD	35.7%
Indeterminate IIP	42.9%
IPAF	3.6%
Pneumoconiosis	3.6%



Adverse Events



Nebulised Pirfenidone appears to be a **safe** and **well-tolerated** treatment in patients with **PPF. Cough** was the most commonly reported **adverse event (AE)** with a rate comparable to placebo. **Gastrointestinal** side-effects are markedly reduced compared with currently licensed treatment for **PPF**.

This first look at the **FVC data** suggests **nebulised Pirfenidone** is a promising development for the treatment of patients with **PPF**.

A Phase 2 study of **nebulised Pirfenidone** in **PPF** is planned and is aiming to recruit the first participant from early 2024.

Conflict of interest disclosure:
Dr A West: Honoraria from Boehringher Ingelheim, (Avalyn Pharma Inc.)
H Bao, D Nair, C Thompson, Dr F Woodhead: Employees of Avalyn Pharma Inc.

References
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