

# TREATMENT OF PULMONARY EMPHYSEMA ASSOCIATED WITH ALPHA-1-ANTITRYPSIN DEFICIENCY

S. Llamas, S. El Mahmoud, E. Gutiérrez, D. López, A. Castillo, J. Juan, J. Ortiz de Urbina, L. Ortega, J. Medina, R. Ruano

## Background

The **alpha-1-antitrypsin deficiency (AATD)** is an inherited disorder that predisposes to the development of emphysema. Replacement therapy is a chronic treatment with a high economic impact.

## Purpose

Describe the characteristics of patients diagnosed with AATD and emphysema in our hospital. Follow-up their evolution through the values of spirometric data and the number of hospitalization basal and after one year treatment.

## Material and methods

A retrospective study of the individual evolution with AATD who have been treated with replacement therapy. Review of medical records and pharmacy dispensing data.

**Variables:** : sex, age at diagnosis, phenotype, FVC, FEV1, FEV1/FVC and number of exacerbations that required hospitalization before and after treatment.

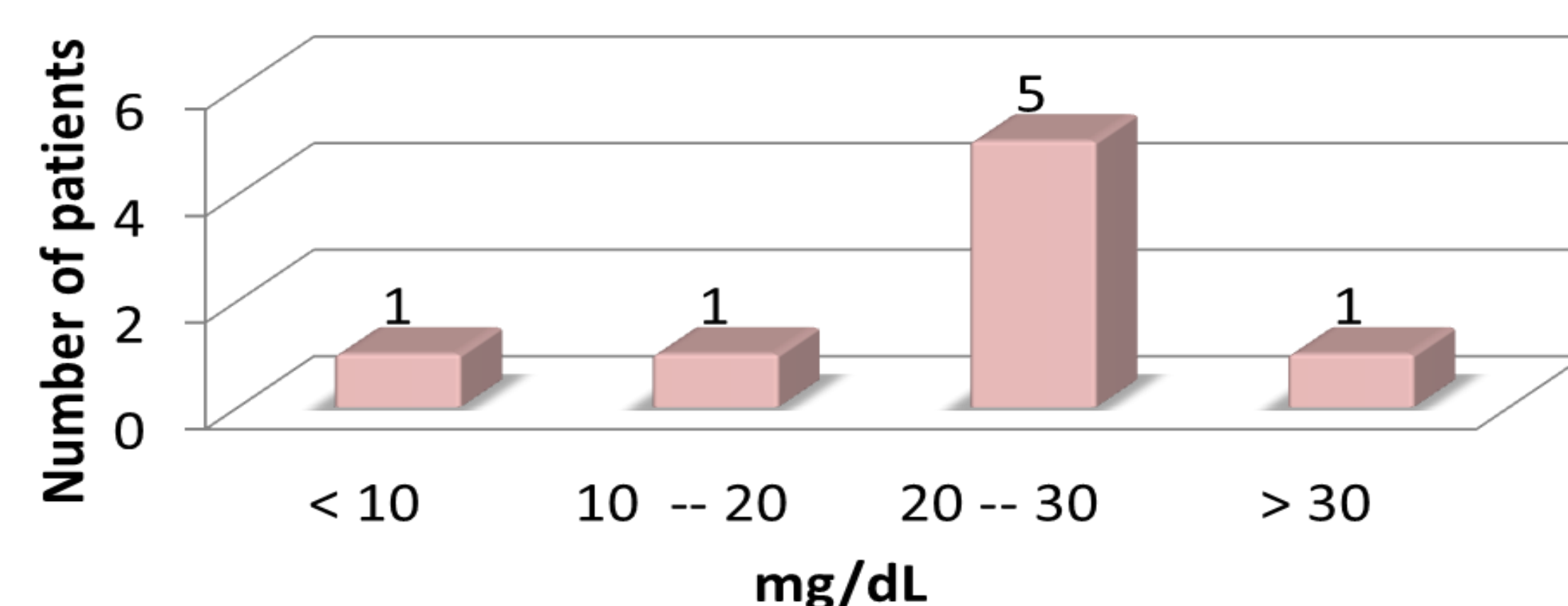
## Results

- 8 patients received replacement therapy.

### Age of diagnosis

Of age	Number of patients	%
< 40	1	12,5
40 – 50	2	25
50 – 60	3	37,5
> 70	1	12,5
$\bar{X}$	51 years	

AAT values at diagnosis



Phenotype	Number of patients	%
piZZ	7	87,5
pi null/null	1	12,5

- In 2 patients, the number of hospitalizations was minor once the treatment started. In 1 patient the number of hospitalizations increased.
- The average **FEV1** before starting treatment was 1.30 0.19L and year after 1.33 0.48L, representing an increase of **0.029L**.
- The average **FEV1/FVC** prior to treatment was 42.9 8.02L and 46.15 7.24L a year after, increasing **3.26L** after one year treatment.

## Conclusions

The objective in AAT replacement therapy is to slow down the progression of emphysema and respiratory functional impairment, reducing severe exacerbations. We might with the results obtained that the treatment is effective because the respiratory parameters in our patients were stable after one year treatment showing a slight increase, and the number of hospitalization due to respiratory causes was practically unchanged. However, the limited number of cases and follow-up is insufficient to generalize findings. Larger cohorts would be required.