

Recommendations for the Implementation of the Self-Administration of Alpha-1 Antitrypsin

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Purpose: Administration of exogenous alpha-1 antitrypsin (AAT) is the only specific therapy for the management of pulmonary morbidity in patients with AAT deficiency. It requires weekly or biweekly intravenous infusions, which may impact patient independence and quality of life. Self-administration of AAT therapy is an alternative to reduce the burden for patients who require AAT therapy. We presented herein experts' recommendations for the implementation of a program for the self-administration of AAT.

Methods: This project was conducted using a modified nominal group technique and was undertaken in two online meetings involving the participation of 25 experts: specialists in pulmonology (n=17), nurses (n=5) and hospital pharmacists (n=3).

Results: The following issues were discussed, and several recommendations were agreed upon on the following topics: a) patient profile and clinical evaluation, establishing selection criteria that should include clinical as well as social criteria; b) role of health care professionals, suggested roles for specialists in pulmonology, nurses, and hospital pharmacists; c) training by the nurse, including recommendations before initiating the training and the content of the training sessions; and d) logistic issues and follow-up, adherence, and patient support.

Conclusion: We expect this proposal to increase awareness of this therapeutic alternative and facilitate the implementation of self-administration programs, thus contributing to optimizing the patient experience with AAT therapy. Further research on the outcomes of these programs, especially from the patient perspective, will also help to improve their design and implementation.

Keywords: alpha-1 antitrypsin deficiency, disease burden, augmentation therapy, self-administration

Introduction

Administration of exogenous alpha-1 antitrypsin (AAT) is the only specific therapy for the management of pulmonary morbidity in patients with AAT deficiency.^{1,2} According to the Spanish registry on AAT deficiency, it is received by two-thirds of the patients with the genotype PI*ZZ,³ although large variations exist across Europe.⁴ Administration of exogenous AAT requires weekly or biweekly intravenous infusions, which may impact patient independence and quality of life.⁵ In a recent survey conducted among 16 patients with AAT deficiency who underwent a home-based administration program of AAT therapy, before starting the program, all patients reported that their augmentation therapy interfered “much” or “very much” with their lives.⁶ In the European Alpha-1 Research Collaboration survey conducted in 26 European countries, in nonreimbursed countries, hospital administration of augmentation therapy was considered by patients/caregivers as one of the most challenging barriers to treatment.⁷ Based on previous experiences with the successful self-administration of other intravenous therapies, such as in patients with hemophilia^{8,9} or hereditary angioedema,^{10–12} there is a growing interest in the self-administration of AAT therapy as an alternative to reduce the burden for patients who require AAT therapy. The initial infusions of AAT must be administered under the supervision of a health professional experienced in the treatment of AAT deficiency. However, for the subsequent infusions, an approved product by the European Medicines Agency for AAT therapy is allowed to be administered by a caregiver or by the patients themselves.¹³ Despite the potential advantages of self-administration and that this practice is supported by experts on the management of this clinical condition,⁴ self-administration of AAT therapy has been scarcely adopted by patients and rarely discussed with them by health care professionals (HCP).¹⁴ To improve this situation, several experts advocate for increasing awareness of the self-administration of AAT therapy through the development of guidelines and training programs for both HCP and patients.^{14,15} The objective of this project was to produce a set of recommendations for the implementation of a program for the self-administration of AAT.

Materials and Methods

This project was conducted using a modified nominal group technique, a qualitative method of consensus that is used when the evidence on a specific topic is very limited. It has been applied to different health care contexts, including practice development and education and training,¹⁶ two characteristics of the program we developed. For the scope of this consensus, self-administration was considered any administration out of the hospital that is undertaken by the patient, partner, or caregiver (ie, a nonhealthcare professional).

The project was initiated by the coordinator (MT) with the support of a sponsor and a research organization with expertise in these methods (see acknowledgments). The coordinator, a pulmonologist, selected a group of 5 experts on the management of AAT deficiency, 4 pulmonologists and a nurse who comprised the scientific committee (MCR, J-LL-C, CM-M, MM, APC). An additional group of 19 experts—the expert committee—was also selected based on their experience with the management of AAT deficiency and included pulmonologists (n=12), nurses (n=4) and hospital pharmacists (n=3).

The project was undertaken in two online meetings. The first meeting took place on June 10, 2021 with the participation of the coordinator and the scientific committee with the objective of presenting the project and the methodology to the scientific committee, establishing the timing, and defining the outline for the content of these recommendations. The second meeting was held on October 19, 2021, with all 25 participants present, including the coordinator and the scientific and expert committees. After a plenary session where the participants were told about the project and the process for that meeting, they were split into 5 groups in virtual rooms to discuss one point each of the index and the outline content ([Supplementary Table 1](#)). Each group was coordinated by a member of the scientific committee, and a member of the research organization acted as a facilitator of the discussion. The coordinator presented the initial content of their section as agreed upon in the first meeting by the scientific committee, and the facilitator asked the following: “What aspects do you consider should be modified in the proposed content that should be included in your section of the document?” The attendees spent several minutes thinking about the question and writing down their thoughts on the topics, which were recorded on a whiteboard. Afterward, the participants’ ideas were discussed and clarified in a group discussion, and where appropriate, similar views were grouped together with the agreement of all participants. Then, the facilitator asked the following: “Based on your expert opinion, which of these agreements do you think

should be included in this section?" Finally, all participants met in a second plenary session where the coordinator of each group presented the agreements of the corresponding group, and the members of the other groups had the opportunity to add their views.

Results

Below, we present a summary of the recommendations agreed upon by the experts, and the program is outlined in Figure 1.

Patient Profile and Clinical Evaluation

To establish the profile of patients who are adequate candidates for self-administration, the panel considers that it is important to bear in mind the objectives of self-administration: 1. to empower the patient to actively manage and control the disease; 2. to promote family/work conciliation and patient independence and, thus, improve treatment adherence; 3. to avoid nosocomial respiratory infections in the context of the COVID-19 pandemic; and 4. to reduce the costs associated with AAT therapy, especially those related to nursing activities.

The selection criteria should include clinical as well as social criteria. From the clinical perspective, patients must meet national or international criteria for receiving augmentation therapy, that in the Spanish setting are those of the REDAAT organization (Spanish Network on AAT deficiency)¹⁷ (Box 1); should show hemodynamic stability; should exhibit psychological stability to ensure adherence to the procedures and treatment; and must have the potential ability to perform canalization. From the social perspective, the patients should express their desire to improve their quality of life and/or to achieve greater independence from the hospital center; they should commit, after appropriate training, to perform the administration in an effective and safe manner, including the aseptic conditions for the administration; they should be able and willing to learn the administration technique; they should be stable from a personal point of view; and they should be compliant with their current treatments.

The experts consider that the following situations preclude the participation of a patient in the self-administration program: not providing informed consent, presence of severe thrombocytopenia (<50,000 platelets/ μ L), being under

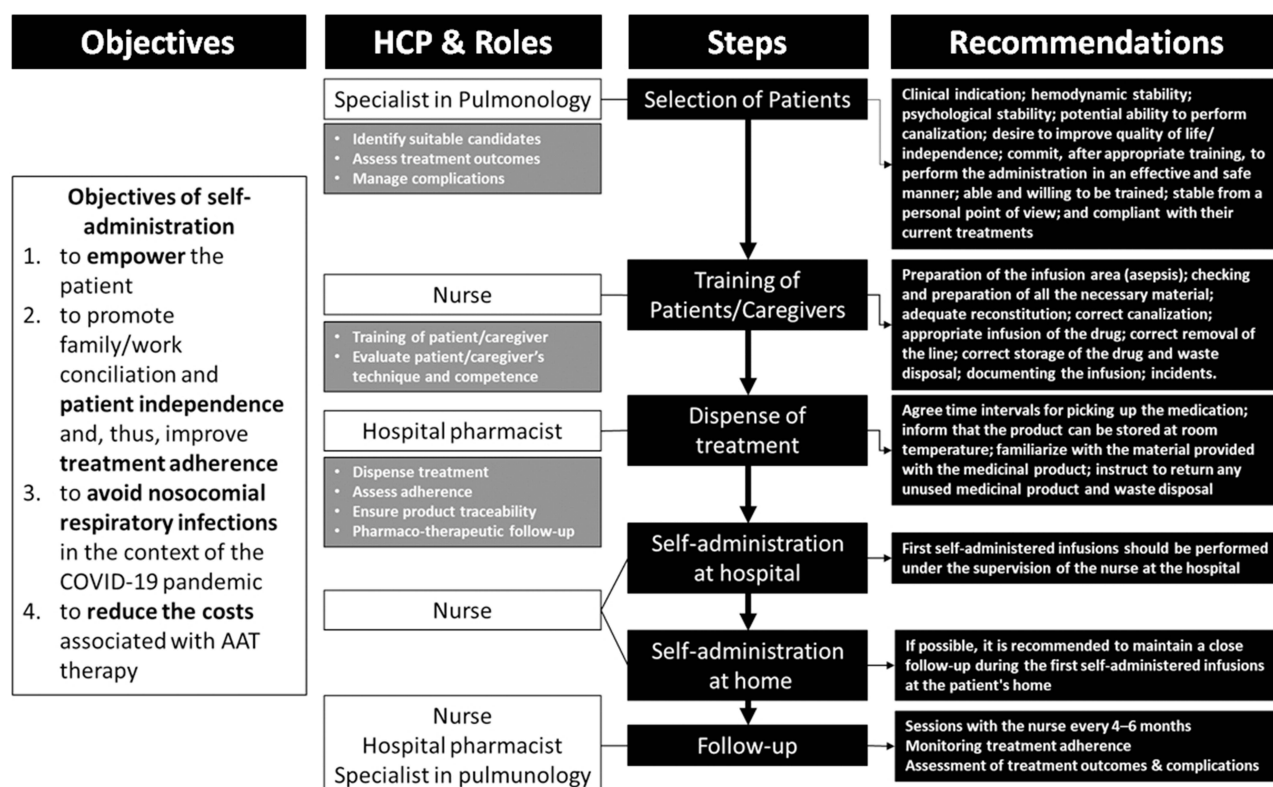


Figure 1 The objectives, steps and recommendations for the implementation of a self-administration program of alpha-1 antitrypsin and the health professionals involved. **Abbreviations:** AAT, alpha-1 antitrypsin; HCP, health care professional.

Box 1 Criteria of the Spanish Network on AAT Deficiency (REDAAT-SEPAR) for the Administration of Alpha-1 Antitrypsin Augmentation Therapy

Age over 18 years
Serious AAT deficiency demonstrated by serum concentrations $\leq 11 \mu\text{M}$ (60 mg/dl)
Nonsmokers or former smokers for at least the last 6 months
Pulmonary emphysema demonstrated by lung function tests and/or chest HRCT
COPD with FEV1 < 80% predicted who receive optimal pharmacological and nonpharmacological treatment
Not presenting an immunoglobulin Ac deficiency
Willing and able to receive regular treatment with AAT i.v. in day hospital

Note: REDAAT-SEPAR, Spanish Network on AAT deficiency- Spanish Society of Pulmonology and Thoracic Surgery (SEPAR).

treatment with anticoagulants, exhibiting New York Heart Association class III or IV heart failure, having a modified Medical Research Council Dyspnea Scale greater than 3 (except when the self-administration is provided by the partner or a caregiver), having a severe mental disorder (eg, a psychotic disorder) or having any social issue that precludes adequate self-administration or the ability to contact a health center.

Role of Health Care Professionals

We consider that HCPs who should be involved in a self-administration program for patients with AAT deficiency are pulmonologists, nurses, and hospital pharmacists. Their respective roles in the self-administration program are presented in [Table 1](#) and can be summarized in three key points: consistent with the summary of product characteristics of the medicinal product, the pulmonologist is responsible for the decision of whether a patient is suitable for self-administration and will ensure that appropriate training is provided; the nurse is responsible for the patient/caregiver's training; and the hospital pharmacist is responsible for dispensing the medicinal product and related issues such as monitoring treatment adherence.

In addition to those roles, the participants consider that patients participating in the self-administration program should keep a diary where they can record information on every self-administered infusion (eg, date of administration, batches, dosage) and potential associated complications, including those related to the administration technique. This diary may help with the follow-up by the HCPs.

To facilitate communication among the HCPs, it is considered that the nurse could complete a checklist with the activities and/or actions related with the self-administration training performed with the patient/caregiver and share it with the hospital pharmacist and the pulmonologist. The proposed checklist is shown in [Box 2](#).

Table 1 Role of Health Care Professionals in a Self-Administration Program

Specialist	Role
Pulmonology	<ul style="list-style-type: none"> Identify the candidate patients and assess their suitability for the program Assess treatment results, treatment satisfaction and adherence Identify complications arising from self-administration and provide potential solutions
Nurse	<ul style="list-style-type: none"> Train the patient/caregiver in self-administration Evaluate patient/caregiver's technique and competence for self-administration.
Hospital pharmacy	<ul style="list-style-type: none"> Dispense treatment Assess adherence Ensure product traceability Pharmaco-therapeutic follow-up

Box 2 Nurse Checklist for the Management of Patients Included in a Self-Administration Program of AAT Therapy

• Has been informed by the responsible pulmonologist.
• Meets clinical and social criteria for inclusion in the self-administration program.
• Has completed and signed the informed consent (this document must be included in the clinical records).
• Has received training in the technique of self-administration and how to perform the treatment safely and effectively.
• After the training process, the patient/caregiver is considered competent to carry out the self-administration of the treatment (this issue must be reflected in the clinical record).
• Has been instructed to fill in the patient's diary correctly.

Training by the Nurse

Although this section focuses on the training of the patients/caregivers, it is important to stress that all HCPs involved in the self-administration program should be trained on the basics of self-administration, including the technique itself and the problems—and how to handle them—that could arise during self-administration.

Potential risks associated with self-administration are related to the handling and administration of the medicinal product as well as to the handling of adverse reactions, particularly hypersensitivity. Therefore, patients/caregivers should be trained in all these aspects. A summary of the key points to be considered for the training of the patients/caregivers is presented in Table 2 and include recommendations before initiating the training and the content of the training sessions; the latter could be used as a checklist to ensure that the patients/caregivers have received adequate training and to inform the other HCPs. First self-administered infusions should be performed under the supervision of the nurse at the hospital; if possible, it is recommended to maintain a close follow-up during the first self-administered infusions at the patient's home. Once the patient starts the self-administration without supervision, it is recommended that follow-up sessions be conducted every 4–6 months to ensure that once the skills have been acquired by a patient/caregiver, they are retained in the long term.

Table 2 Key Points for the Training of the Patients on Self-Administration

Before Initiating the Training	During the Training ^a
Prior consultation, on a different day than that selected for starting the self-administration, so that the responsible nursing staff can resolve doubts and provide the information that the patient may request in advance.	Preparation of the infusion area with an adequate level of asepsis.
Possibility of supervised self-administration in the hospital and, whenever is possible, in the patient's home.	Checking and preparation of all the necessary material.
If possible, evaluation of the patient's home as appropriate setting for self-administration.	Adequate reconstitution of the concentrate.
	Correct canalization.
	Appropriate infusion of the drug.
	Correct removal of the line once the infusion is finished.
	Correct storage of the drug at home and waste disposal (eg, needles).
	Appropriate procedure for documenting the infusion and informing the health professionals responsible of possible incidents arising during self-administration

Note: ^aThese points could be used as a checklist.

In addition to the training on the self-administration technique, the patient/caregiver should receive information on the potential incidents, including the most common adverse events that could arise during self-administration and how to handle them initially. It is also important that hospitals that implement a self-administration program offer support to the patients included in the program through a simple way to contact the HCP for the urgent resolution of doubts or problems with self-administration. In this regard, it is recommended that a pathway for problem solving for these patients be established.

To reinforce the patients/caregivers' competencies for self-administration, it could be useful to provide them with actual patients' testimonies on self-administration, including advantages and potential difficulties.

Logistic Issues

There are several logistic issues that patients/caregivers should be aware of regarding how to pick up the medicinal product, the equipment for the administration, and the disposal of the equipment after self-administration.

Initially, as stated above, medication is dispensed by the hospital pharmacy and, therefore, should be picked up there. The time intervals for picking up the medication should be agreed upon with the hospital pharmacy but should be sufficiently broad to be perceived as an advantage by the patients; however, it is recommended that the intervals not be longer than 3 months. The patients/caregivers should be informed that the medicinal product as provided (ie, before reconstitution) can be stored at room temperature (up to 25 °C).

The patients/caregivers should be familiarized with the material that is provided with the medicinal product: powder vial, solvent vial of water for injections, transfer set for reconstitution, IV infusion set, butterfly set and alcohol swabs.

The hospital pharmacist will instruct the patient/caregivers to return any unused medicinal product and how the waste material should be disposed of and returned. Thus, regular waste (eg, dressing, bags) can be managed normally and disposed of with common garbage; and sharp materials (eg, needles) and material containing biological waste should be disposed of in a sharp container marked with the biohazard symbol, which is resistant to puncture, leak-proof, and safe to handle. In any case, waste material disposal should comply with local regulations and hospital procedures. Appropriate containers will be provided by the hospital pharmacy. It would be useful to prepare some kind of visual aid for patients/caregivers (eg, a table, or infographic) which links the material provided for the self-administration with the type of waste/container that should be used.

Follow-Up, Adherence, and Patient Support

As in other therapeutic areas, treatment adherence and appropriate patient follow-up are essential to ensure the efficacy and safety of AAT therapy.

The monitoring of treatment adherence could be implemented in a complementary way. For example, at the time of dispensing the drug using the corresponding pharmacy registry, which, ideally, could have a red flag system to signal that the patients have not picked up the medication as scheduled; at that time, the pharmacist could ask for empty vials before dispensing the medications; and, as mentioned before, the patient should be encouraged to use a diary to record the medication administered with dates, batch numbers and so on.

It would be useful to prepare a patient resource booklet and/or quick guides providing basic instructions for self-administration, informing about common adverse events and the initial measures to handle them, and including practical recommendations for self-care (eg, skin care). Whenever possible, these resources should be prepared by a multidisciplinary team that includes all stakeholders involved in the self-administration, that is, HCPs and patients. The more visual they are, the more useful they will be for patients.

Patients must be able to clearly identify a HCP of reference, the nurse, who will provide the basic support for self-administration. To facilitate access to this professional, in addition to having a helpline, it is recommended that the patient schedule the infusions with a timing compatible with HCP availability. Patient advocacy groups could also have a role in patients' support.

Discussion

The limited use of self-administration of AAT therapy despite its benefits is probably due to the lack of knowledge of this option by patients, as has been the case with other self-administration programs of parenteral medicines.¹⁸ It is likely that increasing awareness among HCPs about this alternative of administration would lead to better communication between HCPs and the patients and, in turn, would lead to greater awareness and interest among patients with AAT in self-administration. Our program aims to address that issue and to help HCPs implement a self-administration program in their settings. In a US survey among patients participating in the AlphaNet Disease Management and Prevention Program, 44 of the 555 patients who participated in the survey self-administered their AAT infusions, and 95% were very satisfied and 5% were satisfied with their treatment, but no other patient-reported outcomes were provided.¹⁴ Therefore, in our view, it is important to demonstrate the beneficial effects of the self-administration of AAT from the patients' perspective, as it has been shown with other self-administered therapies, such as those for the management of hereditary angioedema.^{19,20}

Another alternative strategy for the administration of AAT therapy is the administration of augmentation therapy at home by nurses. Some authors considered that using nurses for home-based administration has the advantage that they could routinely perform basic health assessments of patients with AAT deficiency.²¹ This strategy was evaluated in 16 patients with AAT deficiency, showing an improvement in the quality of life with no safety issues.⁶ We think that home-based administration by nurses could be an alternative for some patients, but it does not take into consideration the advantages of self-administration, such as self-care competence and independence.^{22,23}

The selection of patients is considered critical for the success of a self-administration program.¹⁵ The criteria we have proposed are based on the opinions of experts. Although they are consistent with other expert recommendations,¹⁵ we should investigate using real-world data which takes in account characteristics of the patients, the disease, and the health care organization that are associated with the success of the program, with success being a complex outcome that should combine adherence to the program, patient's satisfaction and quality of life, efficacy and lack of occurrence of relevant safety issues.

In the above mentioned survey among 44 users of self-administration of AAT therapy, most patients (84%) reported no difficulties with the procedure; the difficulties reported by 5 patients included the choice of the injection site, problems finding a vein, port blockage and intravenous stick injuries.¹⁴ In a qualitative study of potential users of self-administration of intravenous antibiotics, one of the key determinants encouraging patients to engage in self-administrations was the perception of being sufficiently knowledgeable, having adequate skills and feeling competent.¹⁸ A small survey of 22 patients undergoing AAT therapy showed that 8 patients were willing to switch from standard administration to a self-infusion program provided they were trained and educated.²¹ All these data indicate that training the patient is the core of the self-administration program and the area in which we should concentrate our efforts. Regarding the retention of initial training, an experience with self-administration of antibiotics suggests that these training programs could achieve excellent retention.²⁴ However, in our view, it is recommended that follow-up training sessions be conducted to confirm that retention.

A major limitation of our work is that it did not involve the key stakeholder: the patient. We were focused on the organization of the program from the HCP point of view. However, it is essential that, in a second phase, we obtain feedback from patients with AAT deficiency on the materials we have produced and the procedure itself. Another potential limitation is that we have used the term self-administration with a meaning that goes beyond administration by the patients. However, this is consistent with the use of the term by other authors when referring to self-administration of AAT therapy.¹⁵ Finally, health care organizations differ across countries and even within the same country; therefore, our proposal should be customized to each specific setting through, for instance, the development of specific pathways for self-administration. Overall, it is important to stress that this is not a practice guideline, but a set of expert recommendations that needs to be validated. Current studies such as the AmARETI Study²⁵ and future investigations such as those suggested above will help to improve these programs.

Conclusion

Overall, we expect that this proposal increases awareness about this therapeutic alternative and facilitates the implementation of self-administration programs and, thus, contributes to optimizing the patient's experience with AAT therapy.

Abbreviations

AAT, alpha-1 antitrypsin; HCP, health care professionals; REDAAT, Spanish Network on AAT deficiency.

Data Sharing Statement

Not applicable. All the relevant information is presented with the manuscript.

Ethics Approval and Informed Consent

This project did not involve the participation of patients. Due to its nature, following the general Spanish regulations on biomedical research (ie Law 14/2007, of July 3, on Biomedical Research/La Ley 14/2007, de 3 de julio, de Investigación biomédica), this project did not require the evaluation of an Ethics Committee. The participating experts were informed about the nature of the project and agreed to participate by signing a contract.

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Author Contributions

All authors made a significant contribution to the work reported, whether that is in the conception, study design, execution, acquisition of data, analysis and interpretation, or in all these areas; took part in drafting, revising or critically reviewing the article; gave final approval of the version to be published; have agreed on the journal to which the article has been submitted; and agree to be accountable for all aspects of the work.

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