

Individual Health Management Plan

A long-term guide for
Alphas with Lung Disease

Skinny Little Reference Guide™



This is a guide to help build a collaborative plan between you and your healthcare provider. The goal of a health management plan is to look beyond the next scheduled healthcare provider visit and develop a plan to maintain health and evaluate for changes with a view that spans months or years into the future. It is recommended that this collaborative health management plan be reviewed with your healthcare provider on an annual basis.

The column on the right, “My Personal Guide to Alpha-1 Treatment” has been prepared to help you navigate the details of dealing with an Alpha-1 diagnosis. It includes places for you to record essential information, discussion topics to talk about with your doctor and/or nurse and direction about tests, issues and lifestyle decisions.

On the left, you have a guide for your doctor and/or nurse to follow. Plan to meet on at least an annual basis and review this information. Be sure to take along this handy reference and share it with those who are providing your medical care. It is also a good idea to keep a list of any additional questions, as well as a list of all your medications, tucked into this brochure so you will have it on-hand.

If your doctor and/or nurse would like their own copy of the “Healthcare Provider Treatment Plan” they can access the information by visiting the AlphaNet website at www.alphanet.org, and printing a copy. This material is part of AlphaNet's **Big Fat Reference Guide**.

This Plan is the property of:

PATIENT NAME: _____

DATE OF BIRTH: _____

AGE: _____ SEX: _____

DATE OF ALPHA-1 DIAGNOSIS: _____

TODAY'S MONTH/YEAR: _____

HEALTHCARE PROVIDER TREATMENT PLAN

MY PERSONAL GUIDE TO ALPHA-1 TREATMENT



Diagnosis

Alpha-1 Antitrypsin Deficiency

- Confirmed (1 time)
 - Phenotype
 - Genotype
 - Alpha-1 level
- Consider consultation with an Alpha-1 lung or liver specialist
- Genetic counseling and family testing
- Discussion of membership in the Alpha-1 Research Registry and DNA and Tissue Bank
- Discussion of participation in appropriate clinical trials

Pulmonary Diagnosis

- COPD
 - Emphysema
 - Chronic Bronchitis
- Bronchiectasis
- Asthma
- Other: _____

Evaluate for Related Medical Conditions

- Gastroesophageal reflux/aspiration
- Sinusitis
- Liver disease
- Sleep apnea and nocturnal desaturation
- Pulmonary hypertension
- Osteoporosis, bone loss
- Necrotizing panniculitis
- Wegener's granulomatosis
- Atypical mycobacteria
- Rare conditions associated with Alpha-1: _____

Discuss Alpha-1 diagnosis

- Phenotype _____
- Genotype _____
- Alpha-1 level _____
- Ask about evaluation by a healthcare professional with expertise in Alpha-1 (lung and/or liver disease)
- Discuss genetic and hereditary considerations
- Discuss family testing
- Consider participation in research studies

Pulmonary Diagnosis

I'm an Alpha with lung disease

Emphysema _____

Chronic Bronchitis _____

Bronchiectasis _____

Asthma _____

Other: _____

Report the following symptoms:

- Heartburn
- Sinus problems
 - Constant or night time drip/drainage
 - Sore throat
- Sleep problems
 - Snoring
 - Tired when arise
- Skin problems
 - Rash/itching/pain/eruptions
- Liver symptoms
 - Jaundice (skin yellowing)
 - Bleeding

HEALTHCARE PROVIDER

MY PERSONAL GUIDE



Health Status Assessments

(annual, at minimum)

Laboratory

- CBC, platelets
- PT
- ABG's or oximetry
- ALT, AST, GGTP, Bilirubin total and direct, LDH, Alk.Phos.

Pulmonary Function Testing

- Complete pulmonary function tests pre- and post-bronchodilator including plethysmographic lung volumes and DLCO
 - Spirometry alone (if more complete testing unavailable)

6-minute walk with oximetry and titration

Classify COPD severity

- Consider lung transplant evaluation if Very Severe air flow obstruction

Radiology

- Chest PA and lateral or baseline high resolution CT of chest (1 time only) or follow-up CT of chest (if change in clinical status)
- Bone densitometry (baseline and as indicated)

Medications

Review with emphasis on simplification of the regime, new therapeutics and better self-management by patient

Usual medications for Alpha-1 COPD

- Long acting beta-agonist
- Long acting anticholinergic
- Rescue Inhaler
- Augmentation Therapy
 - Prolastin®
 - Zemaria®
 - Aralast™
- 60mg/kg weekly (recommended) over _____ min.

Laboratory

Ask healthcare provider to discuss lab tests and implication for lung status, liver status, and other conditions

Pulmonary Function Testing

Discuss concerns regarding pulmonary function testing

- Difficulties performing test
- Side effects from bronchodilator
- Health status at the time of testing
- Problems withholding pulmonary medications during testing

Discuss status of lung disease and potential for lung transplantation

Radiology

- Discuss concerns regarding radiation exposure
- Discuss results and implication for lung status

Medications

- Bring list of all medications to review with physician
- Review the expiration dates on all home medications
- Discuss side effects/problems
- Ask if medications can be reduced or eliminated
- Keep a comprehensive written list of all medications readily available

I'm on:

Long acting bronchodilator(s) _____

Inhaled steroids _____

Steroid pills _____

Rescue inhaler _____



HEALTHCARE PROVIDER

Medications (continued)

- Infusion schedules other than weekly can be considered

At-home medications prescribed to manage exacerbations/lung infection in the home

- Directions given for home use of:
 - Antibiotics
 - Short course oral steroids
 - Inhaled medications

Patient's trend in exacerbation frequency is:

- No exacerbations
- More frequent or severe exacerbations
- Less frequent or severe exacerbations

Oxygen Prescription

(include justifying ABG/oximetry)

Rest ___ Exertion ___ Sleep ___

MY PERSONAL GUIDE

Medications (continued)

Augmentation Therapy

Brand: _____

Frequency: _____

Dose: _____

During an exacerbation or "flare" of my lung disease:

- I can self-treat at home
 - Drink fluids
 - Sputum clearance techniques
 - Relaxation
 - I start antibiotics and have them at home for this reason
 - I increase my inhaled medications and adjust them to my symptoms as planned with my healthcare provider
 - I use steroid pills and have them at home as planned with my healthcare provider
 - I know when I need to call my healthcare provider, go to the emergency room or call 911
- Discuss issues related to oxygen therapy
- Changes in breathing with rest/exercise/sleep
 - Types of systems/equipment
 - Check insurance coverage

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Toxic Exposure

Assess potential for inhaled toxic exposure in the home and workplace

Assess use of alcohol/other liver toxic substances including medications

Diet and Nutrition

Current Weight: _____

Overweight

- Consider dietary consultation
- Specific diet recommendations
- Recommendations for exercise programs

Home exercise program
Pulmonary Rehabilitation

Underweight

- Dietary consultation with on-going intervention until normal weight restored
- Nutrition plan with consideration of dietary supplements and/or medical nutrition intervention
- Pulmonary Rehabilitation

MY PERSONAL GUIDE

Toxic Exposure

- Discuss the potential for toxic exposure in the workplace with supervisor
- Have strategies in place to avoid occupational dust and fume exposure
- As appropriate, have a properly fitted mask
- Know how to access of MSDS at work

I'm aware of those substances in my home or outdoors that are toxic or irritating to my lungs and have a plan to avoid them

Examples: Cleaning substances/
wood smoke

Discuss the risks associated with alcohol consumption and Alpha-1. Ask about the effects of your medications on the liver

Diet and Nutrition

Current Weight: _____

This weight is:

Overweight _____

Underweight _____

Desired weight _____

Overweight

- Develop or enroll in a weight management plan
- Develop or enroll in an exercise program
- Discuss the use of vitamins and mineral supplements

Underweight

- Discuss the need for a nutrition evaluation with healthcare provider
- Discuss the use of vitamins and mineral supplements and potential need for other nutritional interventions
- Discuss pulmonary rehabilitation and exercise limitations until weight stabilized



Lifestyle Management

Immunizations

- Influenza vaccine
- Pneumococcal vaccine (q 5 yrs. if COPD)
- Hepatitis A vaccine
- Hepatitis B vaccine

Smoking Cessation

- Referral to smoking cessation program
- Nicotine replacement therapy prescribed

Immunizations

- Flu Shot (annual)
- Pneumonia vaccine (every 5 years if COPD)
- Hepatitis A vaccine
- Hepatitis B vaccine

Smoking Cessation

- I don't smoke or have a plan in place to stop
- I'm in a smoking cessation program
- I use my nicotine replacement therapy
- I have a strategy in place to avoid second-hand smoke



HEALTHCARE PROVIDER

Activity and Fitness: Improvement and Maintenance

Assess current fitness level

Home Exercise Program recommendations:

- Warm-up and stretching
- Muscle strengthening
- Cardiopulmonary (endurance)

Pulmonary Rehabilitation referral

- For endurance and strength
- For ADL and pacing
- For 6-minute walk
- For oximetry at rest and with exertion
- For instruction for self-monitoring

Health Insurance

Consider insurance related issues when making referrals, prescribing medications and with other healthcare decisions where cost/reimbursement may significantly impact access to care and services.

MY PERSONAL GUIDE

Activity and Fitness: Improvement and Maintenance

Develop and implement a specific exercise program based on your healthcare provider's recommendation, your motivation and perceived level of fitness

- For weight loss____
- For improved functioning____
- For maintenance____

Home Exercise Program

- Discuss use of oxygen with exercise
- Request specific recommendations for warm-up and stretching, muscle strengthening and cardiopulmonary (endurance) training.
- Date started_____

Discuss the need for a referral to a Pulmonary Rehabilitation Program

- Check insurance coverage
- Date started_____

Health Insurance

I am aware of what my current health insurance provides

- Lifetime maximum
- Major medical
- Disability coverage

I have evaluated insurance options based on my age, employment and income

I have considered appropriate supplemental or secondary insurance

I keep careful records and don't hesitate to question denials or coverage issues

HEALTHCARE PROVIDER

MY PERSONAL GUIDE



Coping and Support Strategies

Assess understanding and acceptance of diagnosis

- Discuss long term implications of diagnosis with chronic disease
- Discuss participation in local support organizations

Assess for presence of Depression

- Consider professional evaluation if symptoms persist or become severe
- Consider antidepressants

Provide opportunity for discussion of issues related to sexuality/sexual performance/dysfunction as related to COPD

- Consider referral to specialist

Discuss medical and lifestyle implications of organ transplantation.

I understand and accept the diagnosis of Alpha-1

My family understands and accepts the diagnosis of Alpha-1

Seek out local and national resources that can be used to support and educate myself and my family

- AlphaNet
- Alpha-1 Association
- Alpha-1 Foundation

Report the following symptoms

- On-going feelings of sadness
- Sleep loss/sleeping excessively
- Chronic fatigue
- Weight loss
- Withdrawal from activities/people
- Thoughts of suicide

Discuss issues of sexuality/sexual performance/dysfunction with your healthcare provider particularly as related to COPD

- Energy requirements/breathing implications/use of oxygen equipment during sexual activity
- Body image

Ask for referrals to appropriate support services

Discuss lung transplantation/preparatory issues, lifestyle implications

- Selecting a program
- Getting listed
- Discuss potential with family
- Seek support from transplanted Alpha's

HEALTHCARE PROVIDER

**End of Life Planning/
Advance Directives**

Explain, ascertain and document patient's advance medical directives

- Living Will
- Identify responsible family member(s) or holder of durable medical power of attorney
- DNR

Discuss organ donation

MY PERSONAL GUIDE

**End of Life Planning/
Advance Directives**

Discuss your end of life wishes with your healthcare provider and family

Insure that your wishes are known and carried out by preparing appropriate documents

- Living Will
- Durable medical power of attorney
- DNR

Consider organ donation



PHYSICIAN SIGNATURE:

DATE:

PATIENT SIGNATURE:

DATE:

This form is provided as an example only. Always consult your healthcare provider for specific instructions about your individual medical treatments.



This brochure is produced by AlphaNet as part of its Alpha-1 Disease Management and Prevention (ADMAP) program.

AlphaNet is a not-for-profit organization providing disease management services and support to individuals affected by Alpha-1 through a staff of medical professionals and specially trained AlphaNet Patient Services Coordinators, available 24 hours a day, 7 days a week. To learn more about ADMAP or to find the AlphaNet Coordinator nearest you, visit our website (www.alphanet.org).