



Lung Disease

INDIVIDUAL HEALTH
MANAGEMENT PLAN



HEALTH MANAGEMENT PLAN FOR LUNG DISEASE

You and your doctor are a team, working together to manage your health for the months or years ahead. This Health Management Plan for Alphas with Lung Disease will help. We recommend that you review your plan together every year.

Using your Health Management Plan

In your Health Management Plan, the column on the right helps you navigate life after an [Alpha-1 lung disease](#) diagnosis. Use it to record:

- Essential information
- Things you want to talk to your doctor or nurse about
- Pre- and post-test instructions
- Lifestyle decisions
- Issues, concerns, and questions

The column on the left is a guide for your doctor or nurse to follow. As we recommended, plan to meet at least once a year to review this information. You can also share your guide with other healthcare providers. And if you'd like, tuck a list of your medicines into it, so you'll have the information close at hand if you need it.

How to get your Health Management Plan

Just [download a PDF](#) and print your plan by visiting www.alphanet.org/living-with-alpha-1 and navigating to the Health Management Plan for Lung Disease Guide. If your doctor or nurse wants their own copy, you can email the PDF to them. Or, you can send them there to download their own copy.

DIAGNOSIS	
Healthcare Provider	Alpha
<p>Alpha-1 Antitrypsin Deficiency Confirmed (1 time)</p> <ul style="list-style-type: none"> • Pi-Type • 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 • Discussion of participation in appropriate clinical trials 	<p>Discuss Alpha-1 diagnosis</p> <ul style="list-style-type: none"> • Pi-type _____ • 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 the Alpha-1 Research Registry and research studies
<p>Pulmonary Diagnosis</p> <p>Alpha-1 Antitrypsin Deficiency</p> <ul style="list-style-type: none"> • COPD <ul style="list-style-type: none"> • Emphysema • Chronic Bronchitis • Bronchiectasis • Asthma • Other: _____ 	<p>Pulmonary Diagnosis</p> <p>I'm an Alpha with lung disease</p> <ul style="list-style-type: none"> • Emphysema _____ • Chronic Bronchitis _____ • Bronchiectasis _____ • Asthma _____ • Other: _____
<p>Evaluate for Related Medical Conditions</p> <ul style="list-style-type: none"> • Gastroesophageal reflux/aspiration • Sinusitis • Liver disease • Sleep apnea and nocturnal desaturation • Pulmonary hypertension • Osteoporosis, bone loss • Necrotizing panniculitis • Granulomatosis with Polyangiitis • Atypical mycobacteria • Rare conditions associated with Alpha-1: _____ 	<p>Report the following symptoms:</p> <ul style="list-style-type: none"> • Heartburn • Sinus problems <ul style="list-style-type: none"> • Constant or nighttime drip/drainage • Sore throat • Sleep problems <ul style="list-style-type: none"> • Snoring • Tired when I wake up • Skin problems <ul style="list-style-type: none"> • Rash/itching/pain/eruptions • Liver symptoms <ul style="list-style-type: none"> • Jaundice (skin yellowing) • Bleeding

HEALTH STATUS ASSESSMENTS (ONCE A YEAR, AT LEAST)	
Healthcare Provider	Alpha
<p>Laboratory</p> <ul style="list-style-type: none"> • CBC, platelets • PT • ABG's or oximetry • ALT, AST, GGTP, Bilirubin total and direct, LDH, Alk.Phos. 	<p>Laboratory</p> <p>Ask healthcare provider to discuss lab tests and implication for lung status, liver status, and other conditions</p>
<p>Pulmonary Function Testing</p> <ul style="list-style-type: none"> • 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 <p>Classify COPD severity</p> <p>Consider lung transplant evaluation if very severe air flow obstruction</p>	<p>Pulmonary Function Testing</p> <p>Discuss concerns regarding pulmonary function testing</p> <ul style="list-style-type: none"> • Difficulties performing test • Side effects from bronchodilator • Health status at the time of testing • Problems withholding pulmonary medications during testing <p>Discuss status of lung disease and potential for lung transplantation</p>
<p>Radiology</p> <ul style="list-style-type: none"> • 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) 	<p>Radiology</p> <ul style="list-style-type: none"> • Discuss concerns regarding radiation exposure • Discuss results and implication for lung status

HEALTH STATUS ASSESSMENTS CONTINUED (ONCE A YEAR, AT LEAST)

Healthcare Provider	Alpha
<p>Medications</p> <p>Medication review with special emphasis on simplification of the medical regimen, new therapeutics on the horizon, and better self-management by patient.</p> <p>Usual medications for Alpha-1-COPD</p> <ul style="list-style-type: none"> • Long acting beta-agonist • Inhaled corticosteroid • Long acting muscarinic • Rescue inhaler <p>At-home medications to manage exacerbations</p> <ul style="list-style-type: none"> • Antibiotic (take at first sign of lung infection): • Short course oral steroids <p>Patient's trend in exacerbation frequency is:</p> <ul style="list-style-type: none"> • No exacerbations • More frequent or severe exacerbations • Less frequent or severe exacerbations <p>Oxygen prescription (including justifying ABG and/or oximetry):</p> <ul style="list-style-type: none"> • Rest _____ • Exertion _____ • Sleep _____ 	<p>Medications</p> <ul style="list-style-type: none"> • Bring list of all medications to review with physician • Review the expiration dates on all home medicines • Discuss side effects/problems • Ask if medicines can be reduced or eliminated <p>Keep a complete written list of all your medicines where you can easily find it.</p> <p>I'm on:</p> <ul style="list-style-type: none"> • Long acting bronchodilator(s) _____ • Inhaled steroids _____ • Steroid pills _____ • Rescue inhaler _____ <p>During a flare-up (exacerbation)</p> <ul style="list-style-type: none"> • I can self-treat at home <ul style="list-style-type: none"> • 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 <p>Discuss issues related to oxygen therapy</p> <ul style="list-style-type: none"> • Changes in breathing with rest/exercise/sleep • Types of systems/equipment • Check insurance Coverage

LIFESTYLE MANAGEMENT

Healthcare Provider	Alpha
<p>Immunizations</p> <ul style="list-style-type: none"> • Influenza vaccine • COVID-19 vaccine • Pneumococcal vaccine (q 5 yrs. if COPD) • Prevnar-13 (for transplantees and discuss after 65 yrs.) • Hepatitis A vaccine • Hepatitis B vaccine • Tdap <p>Smoking Cessation</p> <ul style="list-style-type: none"> • Referral to smoking cessation program • Nicotine replacement therapy prescribed <p>Ancillary Information</p> <ul style="list-style-type: none"> • Consultation with an Alpha-1 Aware healthcare provider or Alpha-1 Clinical Resource Center over coming years • Family/genetic counseling and consideration of family testing for Alpha-1 • Discussion of participation in local support group, the Alpha-1 Research Registry, and the Alpha-1 DNA and Tissue Bank • Discussion of participation in appropriate clinical trials 	<p>Immunizations</p> <ul style="list-style-type: none"> • Influenza vaccine • COVID-19 vaccine • Pneumococcal vaccine (q 5 yrs. if COPD) • Hepatitis A vaccine • Hepatitis B vaccine • Tdap <p>Smoking Cessation</p> <ul style="list-style-type: none"> • 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 <p>Ancillary Information</p> <ul style="list-style-type: none"> • Discuss evaluation by a healthcare provider with special expertise in Alpha-1 • Discuss genetic and hereditary considerations • Discuss risks and benefits of testing family members • Find out about local and national resources that can help me educate myself and help others • Consider learning about and joining in drug studies

LIFESTYLE MANAGEMENT CONTINUED

Healthcare Provider	Alpha
<p>Toxic Exposure</p> <ul style="list-style-type: none"> Assess potential for inhaled toxic exposure in the home and workplace Assess use of alcohol/ other liver toxic substances including medications 	<p>Toxic Exposure</p> <ul style="list-style-type: none"> 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 MSDS at work <p>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</p> <p>Examples: Cleaning substances/ wood smoke</p> <p>Discuss the risks associated with alcohol consumption and Alpha-1. Ask about the effects of your medications on the liver</p>

LIFESTYLE MANAGEMENT CONTINUED

Healthcare Provider	Alpha
<p>Diet and Nutrition</p> <p>Current Weight: _____</p> <p>If Overweight</p> <ul style="list-style-type: none"> Consider dietary consultation Specific diet recommendations Recommendations for exercise programs Home exercise program Pulmonary Rehabilitation <p>If Underweight</p> <ul style="list-style-type: none"> Dietary consultation with on-going intervention until normal weight restored Nutrition plan with consideration of dietary supplements and/or medical nutrition intervention Pulmonary Rehabilitation 	<p>Diet and Nutrition</p> <p>Current Weight: _____</p> <p>This weight is: Overweight _____ Underweight _____ Desired weight _____</p> <p>If Overweight:</p> <ul style="list-style-type: none"> Develop or enroll in a weight management plan Develop or enroll in an exercise program Discuss the use of vitamins and mineral supplements <p>If Underweight:</p> <ul style="list-style-type: none"> 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 CONTINUED

Healthcare Provider	Alpha
<p>Activity and Fitness: Improvement and Maintenance</p> <p>Assess current fitness level</p> <p>Home Exercise Program recommendations:</p> <ul style="list-style-type: none"> • Warm-up and stretching • Muscle strengthening • Cardiopulmonary (endurance) <p>Pulmonary Rehabilitation referral</p> <ul style="list-style-type: none"> • For endurance and strength • For ADL and pacing • For 6-minute walk • For oximetry at rest and with exertion • For instruction for self-monitoring 	<p>Activity and Fitness: Improvement and Maintenance</p> <p>Develop and implement a specific exercise program based on your healthcare provider's recommendation, your motivation and perceived level of fitness for</p> <p>Weight loss _____</p> <p>Improved functioning _____</p> <p>Maintenance _____</p> <p>Home Exercise Program</p> <ul style="list-style-type: none"> • Discuss use of oxygen with exercise • Request specific recommendations for warm-up and stretching, muscle strengthening and cardiopulmonary (endurance) training. <p>Date started _____</p> <p>Discuss the need for a referral to a Pulmonary Rehabilitation Program</p> <ul style="list-style-type: none"> • Check insurance coverage <p>Date started _____</p>

COPING AND SUPPORT STRATEGIES

Healthcare Provider	Alpha
<p>Assess Understanding and Acceptance of Diagnosis</p> <ul style="list-style-type: none"> • Discuss long-term implications of diagnosis with chronic disease • Discuss participation in local support organizations <p>Assess for presence of depression</p> <ul style="list-style-type: none"> • Consider professional evaluation if symptoms persist or become severe • Consider antidepressants <p>Provide opportunity of discussion of issues related to sexuality/sexual performance/dysfunction as related to COPD</p> <ul style="list-style-type: none"> • Consider referral to a specialist <p>Discuss medical and lifestyle implications of organ transplantation</p>	<p>I understand and accept the diagnosis of Alpha-1, and my family does, too.</p> <p>My family understands and accepts the diagnosis of Alpha-1 and I can find local and national resources to support and educate myself and my family</p> <ul style="list-style-type: none"> • Alpha-1 Foundation • AlphaNet <p>Report the following symptoms</p> <ul style="list-style-type: none"> • 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 <p>Ask for referrals to appropriate support services</p> <p>Discuss lung transplantation/preparatory issues, lifestyle implications</p> <ul style="list-style-type: none"> • Selecting a program • Getting listed • Discuss potential with family • Seek support from transplanted Alphas



Thank you to our sponsors:

GRIFOLS

CSL Behring
Biotherapies for Life™



This single topic brochure is one of a series extracted from AlphaNet's Big Fat Reference Guide to Alpha-1 (the BFRG), which is available on the AlphaNet website (www.alphanet.org).

To find the AlphaNet Coordinator nearest you, visit our website at www.alphanet.org.

Copyright © AlphaNet, Inc. 2021