

### Diagnosis

#### Alpha-1 Antitrypsin Deficiency

Confirmed (1 time)

- 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

#### 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 the Alpha-1 Research Registry and research studies

#### Pulmonary Diagnosis

Alpha-1 Antitrypsin Deficiency

- COPD
  - Emphysema
  - Chronic Bronchitis
- Bronchiectasis
- Asthma
- Other: \_\_\_\_\_

#### Pulmonary Diagnosis

I'm an Alpha with lung disease

- 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
- Granulomatosis with Polyangiitis
- Atypical mycobacteria
- Rare conditions associated with Alpha-1: \_\_\_\_\_

#### Report the following symptoms:

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

### Health Status Assessments

(Once a year, at least)

#### Laboratory

- CBC, platelets
- PT
- ABG's or oximetry

#### Laboratory

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

<ul style="list-style-type: none"> <li>• ALT, AST, GGTP, Bilirubin total and direct, LDH, Alk.Phos.</li> </ul>	
<p><b>Pulmonary Function Testing</b></p> <ul style="list-style-type: none"> <li>• Complete pulmonary function tests pre- and post-bronchodilator including plethysmographic lung volumes and DLCO</li> <li>• Spirometry alone (if more complete testing unavailable)</li> <li>• 6-minute walk with oximetry and titration</li> </ul> <p>Classify COPD severity</p> <p>Consider lung transplant evaluation if very severe air flow obstruction</p>	<p><b>Pulmonary Function Testing</b></p> <p>Discuss concerns regarding pulmonary function testing</p> <ul style="list-style-type: none"> <li>• Difficulties performing test</li> <li>• Side effects from bronchodilator</li> <li>• Health status at the time of testing</li> <li>• Problems withholding pulmonary medications during testing</li> </ul> <p>Discuss status of lung disease and potential for lung transplantation</p>
<p><b>Radiology</b></p> <ul style="list-style-type: none"> <li>• 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)</li> <li>• Bone densitometry (baseline and as indicated)</li> </ul>	<p><b>Radiology</b></p> <ul style="list-style-type: none"> <li>• Discuss concerns regarding radiation exposure</li> <li>• Discuss results and implication for lung status</li> </ul>
<p><b>Medications</b></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"> <li>• Long acting beta-agonist</li> <li>• Inhaled corticosteroid</li> <li>• Long acting muscarinic</li> <li>• Rescue inhaler</li> </ul> <p>At-home medications to manage exacerbations</p> <ul style="list-style-type: none"> <li>• Antibiotic (take at first sign of lung infection):</li> <li>• Short course oral steroids</li> </ul>	<p><b>Medications</b></p> <ul style="list-style-type: none"> <li>• Bring list of all medications to review with physician</li> <li>• Review the expiration dates on all home medicines</li> <li>• Discuss side effects/problems</li> <li>• Ask if medicines can be reduced or eliminated</li> </ul> <p>Keep a complete written list of all your medicines where you can easily find it</p> <p>I'm on:</p> <p>Long acting bronchodilator(s) _____</p> <p>Inhaled steroids _____</p> <p>Steroid pills _____</p> <p>Rescue inhaler _____</p> <p>During a flare-up (exacerbation)</p> <ul style="list-style-type: none"> <li>• I can self-treat at home</li> </ul>

<p>Patient's trend in exacerbation frequency is:</p> <ul style="list-style-type: none"> <li>• No exacerbations</li> <li>• More frequent or severe exacerbations</li> <li>• Less frequent or severe exacerbations</li> </ul> <p>Oxygen prescription (including justifying ABG and/or oximetry):</p> <ul style="list-style-type: none"> <li>• Rest_____</li> <li>• Exertion_____</li> <li>• Sleep_____</li> </ul>	<ul style="list-style-type: none"> <li>- Drink fluids</li> <li>- Sputum clearance techniques</li> <li>- Relaxation</li> <li>• I start antibiotics and have them at home for this reason</li> <li>• I increase my inhaled medications and adjust them to my symptoms as planned with my healthcare provider</li> <li>• I use steroid pills and have them at home as planned with my healthcare provider</li> <li>• I know when I need to call my healthcare provider, go to the emergency room or call 911</li> </ul> <p>Discuss issues related to oxygen therapy</p> <ul style="list-style-type: none"> <li>• Changes in breathing with rest/exercise/sleep</li> <li>• Types of systems/equipment</li> <li>• Check insurance Coverage</li> </ul>
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### Lifestyle Management

<p><b>Immunizations</b></p> <ul style="list-style-type: none"> <li>• Influenza vaccine</li> <li>• Pneumococcal vaccine (q 5 yrs. if COPD)</li> <li>• Pevnar-13</li> <li>• Hepatitis A vaccine</li> <li>• Hepatitis B vaccine</li> <li>• TDaP</li> </ul>	<p><b>Immunizations</b></p> <ul style="list-style-type: none"> <li>• Influenza vaccine</li> <li>• Pneumococcal vaccine (q 5 yrs. if COPD)</li> <li>• Pevnar-13</li> <li>• Hepatitis A vaccine</li> <li>• Hepatitis B vaccine</li> <li>• TDaP</li> </ul>
<p><b>Smoking Cessation</b></p> <ul style="list-style-type: none"> <li>• Referral to smoking cessation program</li> <li>• Nicotine replacement therapy prescribed</li> </ul>	<p><b>Smoking Cessation</b></p> <ul style="list-style-type: none"> <li>• I don't smoke or have a plan in place to stop</li> <li>• I'm in a smoking cessation program</li> <li>• I use my nicotine replacement therapy</li> <li>• I have a strategy in place to avoid second-hand smoke</li> </ul>
<p><b>Ancillary Information</b></p> <ul style="list-style-type: none"> <li>• Consultation with an Alpha-1 Aware healthcare provider or Alpha-1 Clinical Resource Center over coming years</li> <li>• Family/genetic counseling and consideration of family testing for Alpha-1</li> </ul>	<p><b>Ancillary Information</b></p> <ul style="list-style-type: none"> <li>• Discuss evaluation by a healthcare provider with special expertise in Alpha-1</li> <li>• Discuss genetic and hereditary considerations</li> <li>• Discuss risks and benefits of testing family members</li> </ul>

- Discussion of participation in local support group and membership in the Alpha-1 Association, the Alpha-1 Research Registry, and the Alpha-1 DNA and Tissue Bank
- Discussion of participation in appropriate clinical trials

- Find out about local and national resources that can help me educate myself and help others
- Consider learning about and joining in drug studies

**Toxic Exposure**

- Assess potential for inhaled toxic exposure in the home and workplace
- Assess use of alcohol/other liver toxic substances including medications

**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: \_\_\_\_\_

If Overweight

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

Home exercise program

Pulmonary Rehabilitation

**Diet and Nutrition**

Current Weight: \_\_\_\_\_

This weight is:

Overweight \_\_\_\_\_

Underweight \_\_\_\_\_

Desired weight \_\_\_\_\_

If Overweight:

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

<p><u>If Underweight</u></p> <ul style="list-style-type: none"> <li>• Dietary consultation with on-going intervention until normal weight restored</li> <li>• Nutrition plan with consideration of dietary supplements and/or medical nutrition intervention</li> <li>• Pulmonary Rehabilitation</li> </ul>	<p><u>If Underweight:</u></p> <ul style="list-style-type: none"> <li>• Discuss the need for a nutrition evaluation with healthcare provider</li> <li>• Discuss the use of vitamins and mineral supplements and potential need for other nutritional interventions</li> <li>• Discuss pulmonary rehabilitation and exercise limitations until weight stabilized</li> </ul>
<p><b>Activity and Fitness: Improvement and Maintenance</b></p> <p>Assess current fitness level</p> <p>Home Exercise Program recommendations:</p> <ul style="list-style-type: none"> <li>• Warm-up and stretching</li> <li>• Muscle strengthening</li> <li>• Cardiopulmonary (endurance)</li> </ul> <p>Pulmonary Rehabilitation referral</p> <ul style="list-style-type: none"> <li>• For endurance and strength</li> <li>• For ADL and pacing</li> <li>• For 6-minute walk</li> <li>• For oximetry at rest and with exertion</li> <li>• For instruction for self-monitoring</li> </ul>	<p><b>Activity and Fitness: Improvement and Maintenance</b></p> <p>Develop and implement a specific exercise program based on your healthcare provider's recommendation, your motivation and perceived level of fitness</p> <ul style="list-style-type: none"> <li>• For weight loss _____</li> <li>• For improved functioning_____</li> <li>• For maintenance_____</li> </ul> <p>Home Exercise Program</p> <ul style="list-style-type: none"> <li>• Discuss use of oxygen with exercise</li> <li>• Request specific recommendations for warm-up and stretching, muscle strengthening and cardiopulmonary (endurance) training.</li> <li>• Date started _____</li> </ul> <p>Discuss the need for a referral to a Pulmonary Rehabilitation Program</p> <ul style="list-style-type: none"> <li>• Check insurance coverage</li> <li>• Date started _____</li> </ul>
<p><b>Coping and Support Strategies</b></p>	
<p><b>Assess Understanding and Acceptance of Diagnosis</b></p> <ul style="list-style-type: none"> <li>• Discuss long-term implications of diagnosis with chronic disease</li> <li>• Discuss participation in local support organizations</li> </ul> <p>Assess for presence of depression</p>	<p><b>I understand and accept the diagnosis of Alpha-1, and my family does, too.</b></p> <p>My family understands and accepts the diagnosis of Alpha-1</p> <p>I can find local and national resources to support and educate myself and my family</p> <ul style="list-style-type: none"> <li>• Alpha-1 Foundation</li> <li>• AlphaNet</li> </ul>

<ul style="list-style-type: none"> <li>• Consider professional evaluation if symptoms persist or become severe</li> <li>• Consider antidepressants</li> </ul> <p>Provide opportunity of discussion of issues related to sexuality/sexual performance/dysfunction as related to COPD</p> <ul style="list-style-type: none"> <li>• Consider referral to a specialist</li> </ul> <p>Discuss medical and lifestyle implications of organ transplantation</p>	<p>Report the following symptoms</p> <ul style="list-style-type: none"> <li>• On-going feelings of sadness</li> <li>• Sleep loss/sleeping excessively</li> <li>• Chronic fatigue</li> <li>• Weight loss</li> <li>• Withdrawal from activities/people</li> <li>• Thoughts of suicide</li> <li>• Discuss issues of sexuality/sexual performance/dysfunction with your healthcare provider particularly as related to COPD</li> <li>• Energy requirements/breathing implications/use of oxygen equipment during sexual activity</li> <li>• Body image</li> </ul> <p>Ask for referrals to appropriate support services</p> <p>Discuss lung transplantation/preparatory issues, lifestyle implications</p> <ul style="list-style-type: none"> <li>• Selecting a program</li> <li>• Getting listed</li> <li>• Discuss potential with family</li> <li>• Seek support from transplanted Alpha's</li> </ul>
<p><b>End of Life/Advance Directives</b></p> <p>Explain, ascertain and document patient's advance medical directives</p> <p>Discuss organ donation</p>	<p><b>End of life/Advance Directives</b></p> <p>Discuss end of life issues with my healthcare provider and family</p> <p>Ensure that your wishes are known and carried out by preparing appropriate documents</p> <p>Consider organ donation</p>