

## HEALTH CARE PROVIDER TREATMENT PLAN

## MY PERSONAL GUIDE TO ALPHA-1 LIVER DISEASE

### 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 an available Alpha-1 Research clinical trial
- Discussion of participation in appropriate clinical trials

#### Discuss Alpha-1 diagnosis

Pi-Type \_\_\_\_\_

Genotype \_\_\_\_\_

Alpha-1 level \_\_\_\_\_

- Ask about evaluation by a healthcare professional with expertise in Alpha-1
- Discuss genetic and hereditary considerations
- Discuss family testing
- Consider participation in research studies

#### Liver Diagnosis AAT-related Liver Disease

- Exclude other liver diseases
- Evaluate for associated conditions
- Jaundice  
Portal Hypertension  
Esophageal varices  
Bleeding abnormalities  
Ascites  
Hypersplenism  
Encephalopathy  
Renal Insufficiency (hepatorenal syndrome)
- Referral to Hepatologist to monitor liver function
  - Consider Liver Transplant evaluation if signs of liver failure or Portal Hypertension complications

#### Liver Diagnosis I'm an Alpha with Liver Disease

#### Report the following liver symptoms

- Swelling of the abdomen
- Changes in mental acuity
- Diarrhea/vomiting
- Unusual bleeding/difficulty stopping bleeding
- Yellowing of the skin/eyes
- Itching
- Fatigue

Ask about seeing a liver specialist

Discuss status of liver disease and potential for liver transplant

#### Evaluate for AAT Related Medical Conditions

- COPD
- Gastroesophageal reflux/aspiration
- Necrotizing panniculitis
- Granulomatosis with Polyangiitis
- Atypical mycobacteria
- Rare conditions associated with Alpha-1:  
\_\_\_\_\_

#### Report the following symptoms:

- Changes in ease of breathing at rest/with exertion/with sleep
- Heartburn
- Skin problems like rash, itching, pain, pimples

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### Health Status Assessments

(Annual, at minimum)

#### Laboratory

- CBC, platelets
- Albumin
- Vitamin A, D, E, K
- PT, PTT
- ALT, AST, GGTP, Bilirubin total and direct, LDH, Alk.Phos.
- Alpha-Fetoprotein
- Electrolytes, iron
- BUN, Creatinine
- Amylase/lipase
- Hepatitis A, B, C
- HIV
- Ammonia level, if indicated

#### Laboratory

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

#### Pulmonary Function Testing

- Complete pulmonary function tests pre- and post-bronchodilator including plethysmographic lung volumes and DLCO (baseline)
  - Spirometry alone (if more complete testing unavailable or on annual visits)
- 6-minute walk with oximetry and titration

#### Pulmonary Function Testing

##### Discuss concerns regarding pulmonary function testing

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

#### 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)
- Consider Abdominal Ultrasound
- Consider Endoscopy/ERCP
- Consider Liver Biopsy

#### Radiology

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

#### Medications

- Medication review with special emphasis on simplification of the medical regimen, new therapeutics on the horizon, and better self-management by patient

#### Medications

- Bring list of all medications to review with physician
- Review the expiration dates on all home medications
- Discuss side effects or issues associated with specific medications including over the counter

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- Review the liver implications for specific medications, including OTC medications/vitamins/minerals/ dietary supplements

## MY PERSONAL GUIDE TO ALPHA-1 LIVER DISEASE

medications, vitamins/minerals and dietary supplements

- Ask if medications can be reduced or eliminated
- Keep a comprehensive written list of all medications readily available

List your medications here:

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### Lifestyle Management

#### Immunizations

- Influenza vaccine
- Pneumococcal vaccine (q 5 yrs. if COPD)
- Prevnar-13
- Hepatitis A vaccine
- Hepatitis B vaccine
- TDaP

#### Immunizations

- Flu shot (annual)
- Pneumococcal vaccine (q 5 yrs. if COPD)
- Prevnar-13
- Hepatitis A vaccine
- Hepatitis B vaccine
- TDaP

#### Smoking Cessation

- Referral to smoking cessation program
- Nicotine replacement therapy prescribed

#### 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

#### Diet and Nutrition

Identify specific diet recommendations based on type/severity of liver disease symptoms

- Consider dietary consultation
- Protein intake/restrictions

#### Diet and Nutrition

Discuss specific dietary requirements/restrictions

- Ask about seeing a dietitian

## HEALTH CARE PROVIDER TREATMENT PLAN

- Carbohydrate intake/restriction
- Fat intake/restriction
- Frequent small meals
- Vitamin/mineral supplements
- Sodium/fluid restrictions
- Caffeine restriction

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

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

### If Overweight

- Discuss implications of obesity and liver disease
- Consider Dietary Consultation for weight reduction

### Recommendations for exercise programs

- Home exercise program
- Pulmonary Rehabilitation

### If Underweight

- Determine/correct underlying cause
- Dietary consultation with on-going intervention until normal weight restored
- Nutrition plan with consideration of dietary supplements and/or medical nutrition intervention
- Discuss need for physical rehabilitation/exercise program

### **Activity and Fitness: Improvement and Maintenance**

Assess current fitness level with potential limitations based on liver symptom severity

- Fatigue: Morning exercise
- Weight training with cirrhosis
- No high weights; use lower weights with more reps

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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 effect of being overweight on your liver

### If 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 exercise limitations until weight stabilized

### **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 \_\_\_\_\_

## HEALTH CARE PROVIDER TREATMENT PLAN

Home Exercise Program recommendations:

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

Consider Rehabilitation referral

- For endurance and strength
- For ADL and pacing
- For instruction for self-monitoring

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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 Professional Rehabilitation Program

- Check insurance coverage

- Date started \_\_\_\_\_

## 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 of discussion of issues related to sexuality/sexual performance/dysfunction as related to liver symptoms

- Consider referral to a 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 Foundation
- American Liver 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

HEALTH CARE PROVIDER TREATMENT PLAN	MY PERSONAL GUIDE TO ALPHA-1 LIVER DISEASE
	<ul style="list-style-type: none"> <li>• Energy requirements/breathing</li> <li>• Body image</li> </ul> <p>Ask for referrals to appropriate support services</p> <p>Discuss potential for liver transplantation and preparatory issues</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> <ul style="list-style-type: none"> <li>• Explain, ascertain and document patient's advance medical directives</li> </ul> <p>Discuss organ donation</p>	<p><b>End of life/Advance Directives</b></p> <ul style="list-style-type: none"> <li>• Discuss end of life issues with my healthcare provider and family</li> <li>• Ensure that your wishes are known and carried out by preparing appropriate documents</li> </ul> <p>Consider organ donation</p>