## Intravenous augmentation therapy with alpha-1-antiprotease for severe AAT deficiency* Pretreatment testing

- Spirometry with DLCO.
- Liver function tests.
- Serum IgA level.

## Protocol for augmentation therapy

- Product: Pooled plasma-derived alpha-1-antiprotease (Aralast NP, Prolastin-C, Zemaira, and Glassia).
- Interval: Weekly
- Patient's weight: _____kg.
- Calculate patient's dose: 60 mg × weight in kg = _____weekly dose.
- Aralast NP, Prolastin-C, and Zemaira are supplied as lyophilized preparations and require reconstitution according to the package insert. After reconstitution, pooled AAT should be used within three hours.
- Infusion rate depends on specific products used and ranges from ≤0.2 mL/kg/minute to 0.08 mL/kg/minute. Consult package insert. The rate can be adjusted, if needed for patient comfort.

## Supportive care

- Cessation of smoking and avoidance of passive smoke exposure.
- Avoidance of respiratory irritants.
- Pulmonary rehabilitation if reduced exercise capacity.
- Oxygen therapy (as needed).
- Nutritional support as appropriate to maintain healthy body weight.
- Influenza and pneumococcal vaccinations.
- Treatment of respiratory infections (eg, influenza, bacterial bronchitis, flares of bronchiectasis, pneumonia).
- Inhaled bronchodilators and glucocorticoids per guidelines for COPD.
- Vaccination against hepatitis A and B viruses, if not already immune.

AAT: alpha-1 antitrypsin; DLCO: diffusing capacity for carbon monoxide.

* An AAT serum level <11 micromol/L or <57 mg/dL is considered severe deficiency and is usually associated with a genetic variant, such as PI*ZZ or PI*Null. For information about selection of patients for AAT augmentation, refer to UpToDate review on treatment of AAT deficiency.

Infusion of alpha-1-antiprotease can be performed at home after appropriate training or in an infusion center. Epinephrine should be available, if needed.

The US Food and Drug Administration approved regimen is 60 mg/kg, administered weekly. In special circumstances, 120 mg/kg is administered biweekly, or 250 mg/kg is administered monthly.

Vaccination against hepatitis A and B can help prevent superimposed insults to the liver, but is not necessary prior to AAT augmentation therapy, due to the low risk of transmission.