What is Congenital Alpha-1 Proteinase Inhibitor Deficiency (Alpha-1 PI)?

Alpha-1 PI is a protein, made in the liver, that protects the lungs and other tissues from damage. When the body does not make enough protein, the lungs become damaged and you have trouble with many colds and infections, shortness of breath and regular physical activities. Many alpha-1 patients have ongoing issues with bronchitis and pneumonia, and also may develop emphysema.

There are two kinds of emphysema:

**ACQUIRED:** This type of emphysema is usually found in people who have a history of smoking. If the person is not alpha-1 deficient, even though they have emphysema, alpha-1 therapy will not be an appropriate treatment.

**CONGENITAL or HEREDITARY:** For a person who is alpha-1 deficient, the liver does not make enough of the protein because of a defective gene. Alpha-1 therapy can help these patients. This treatment is not a cure for the disease, but can provide the body with the missing protein it needs to protect the lungs and to improve quality of life. The missing protein can be supplied through a product made from plasma, which is the fluid in your blood vessels that carries the many different parts of blood. This product is called “plasma-derived” because it is made by taking the alpha protein inhibitor out of plasma.

**FIRST DOSE:** Your first dose of alpha-1 therapy may be given to you in a medical setting such as a hospital, doctor’s office or clinic. The first dose can also be given in your home by a nurse.

**ACCESS DEVICE:** The nurse will place a catheter in your hand or arm vein. The medication will be given through this catheter. The medicine should take about thirty minutes to infuse through the catheter. After the infusion is completed, the nurse will remove the catheter — and your therapy is complete for the week!
Infusing Alpha-1 Therapy

In many cases, a nurse will infuse your alpha-1 therapy. If you choose to become independent with your medication administration, your nurse will teach you or your caregiver how to place your own catheter and how to infuse your medication. In this case, it is recommended that you have an adult caregiver present who can learn to provide this therapy and place your catheter for you. They must be able to stay with you during the entire infusion. Until all these skills are learned, the nurse will continue to perform these tasks for you.

In the case that you will be learning to start and infuse your own therapy, you will perform the following steps:

**Step 1** is to *make sure your work area is very clean*. Wash your hands thoroughly with soap and water for at least 3 minutes.

**Step 2** is *choosing a vein*. You will be using a tourniquet to help you choose the best vein to use when inserting the needle.

**Step 3** is called *venipuncture*. Venipuncture is the term used to describe putting a needle into the vein you have chosen in order to give the alpha-1 therapy.

**Step 4** is *mixing your medicine*. This involves mixing the concentrate, which is usually a powder, with a special kind of water called the *diluent* (dil-you-ent).

**Step 5** is *injecting your therapy mixture* into another container with fluid before infusing it. You will attach tubing that has a regulator on it to control the flow, or by using a pump that can push the medicine through the catheter. With some therapies, you will also be connecting an extension set that contains a filter. Your nurse will let you know which therapy requires this step.

The final step is to *remove the catheter* from the vein.

Some Alpha-1 medications are not stable for long periods of time and should not be prepared until the catheter is in a vein and secure. The medication must be used within three hours of being mixed or it loses its effectiveness. Other teaching sheets provided will help you learn the steps you need to know to give yourself the alpha-1 therapy.
Some people experience mild side effects from the medication. Any changes in your condition should be reported to your doctor, nurse or pharmacist. Symptoms are not common, but some patients have reported that after receiving alpha-1 therapy, they experience:

- Chills
- Dizziness or lightheadedness
- Fever up to 102°F which may occur anytime within 12 hours after you have your dose. The fever should be gone within 24 hours. If it continues, please notify your doctor.

It is important to tell your doctor if you have any allergies, if you have had reactions to other plasma products before or if you are pregnant. It is important to let your pharmacist and doctor know the names and doses of all the medications you are taking including prescription drugs, over-the-counter medicines or herbal remedies.

Your medication may be delivered to your home. If a nurse is coming to place the IV catheter and infuse your medication, he/she may bring the medication with them. Once you receive your medication, it is important to follow the storage and handling instructions provided on the medication label. Aralast NP, Zemaira® and Glassia™ should be stored at room temperature.

Before you begin, it is important to inspect your medication prior to use. Make sure all protective covers are intact and the solution is clear and colorless after mixing. Check the medication label for your name and the expiration date. Contact your Coram nurse or pharmacist if you notice any problems with the medication or label.

Once you enter the vial(s), the medication **must be infused within three hours.** It is important that you have an IV catheter in place to prevent waste of this medication.

Because the replacement therapy is not the same temperature as your body temperature, you may find that resting your arm on a heated pad or washcloth will keep you from feeling a chill.

If you have any further questions after reading your patient education tools, please contact your pharmacist, nurse or physician.