Alpha-1 Antitrypsin Deficiency and Augmentation Therapy
Prolastin-C Liquid

Dear Nursing Colleague,

Thank you for caring for individuals affected by Alpha-1 Antitrypsin Deficiency (Alpha-1) who are receiving augmentation therapy. Alpha-1 is one of the most common serious genetic disorders and it is believed that less 10,000 of the estimated 100,000 Americans with Alpha-1 have been diagnosed to date. An additional 20 million Americans are estimated to be genetic carriers of this disorder. Alpha-1 can cause liver disease in children or severe liver and lung disease in adults, presenting as early emphysema and cirrhosis. Currently, the World Health Organization, the American Thoracic Society, and the European Respiratory Society, and the Medical and Scientific Advisory Committee of the Alpha-1 Foundation recommend that all adults with the following conditions be tested for Alpha-1COPD including chronic bronchitis and emphysema, chronic asthma, unexplained bronchiectasis, family history of Alpha-1 and chronic liver disease.

This packet contains an instructional DVD along with supplemental written materials comprising an educational program titled "Alpha-1 Antitrypsin Deficiency and Augmentation Therapy". **This program has been created by AlphaNet, Inc., a non-profit health management organization that provides a wide range of support services to individuals with Alpha-1. AlphaNet has created this program as a service to the Alpha-1 community and for the individuals, agencies and facilities that provide infusion services for them.**

The goal is to improve the knowledge of the nurses caring for people with Alpha-1 Antitrypsin Deficiency, including the safe and effective augmentation therapy with Prolastin- C Liquid.

Instructions for this program are included in the packet. Upon successful completion of the Post Test, with a grade of at least 80%, along with the submission of the Registration and Evaluation Forms, each participant will receive a certificate. The expiration date of this course is February 12, 2021.

This continuing nursing educational activity was approved by the Colorado Nurses Association, an accredited by the American Nurses Credentialing Center's Commission on Accreditation.

Planners and presenters of this CNE activity have disclosed no conflict of interest including no relevant financial relationships with any commercial companies pertaining to this CNE activity.

Please don't hesitate to contact AlphaNet should have any questions regarding this educational program.

Sincerely,

Teresa Kitchen, BSN, RN
Clinical Nurse Manager, AlphaNet, Inc

Alpha-1 Antitrypsin Deficiency and Augmentation Therapy
Prolastin-C Liquid

Packet Contents:

I. **Left File Pocket**

1. Program Flash Drive or can be viewed on line at www.alphanetprofessionaled.org
2. Registration Form
3. Post Test Answer Sheet
4. Program Evaluation
5. Contact Information
6. Fax Cover Sheet

II. **Center Binder Section**

1. Guidelines for Home Intravenous Prolastin-C Liquid Infusion
2. Guidelines for Prolastin-C Liquid Home Mixing-Syringe Method
3. Prolastin-C Liquid Infusion Check List
4. Nursing Care Plan-Augmentation Therapy with Prolastin-C Liquid
5. Prolastin-C Liquid Package Insert Grifols Biotherapeutics
6. Post Test

III. **Right File Pocket**

1. "A Healthcare Provider's Guide to Alpha-1 Antitrypsin Deficiency"

Instructions:

1. View the flash drive presentation and review the written materials. Additional copies of the booklet "A Healthcare Provider's Guide to Alpha-1 Antitrypsin Deficiency" may be viewed or downloaded at the Alpha-1 Foundation's website www.alpha1.org. Go to "Publications" then click on the link to download a copy. Extra copies may also be requested directly from the Alpha-1 Foundation at 877-228-7321.

3. Each participant must complete and submit: (copies of each form should be made as needed)
   - **Registration Form**
   - **Post Test Answer Sheet**
   - **Program Evaluation**

4. Fax the completed forms to:
   Teresa Kitchen BSN, RN
   Clinical Nurse Manager
   AlphaNet, Inc.
   Fax: 1-618-659-4867

Follow Up

All tests will be reviewed and corrected – a minimum grade of 80% will be considered passing. A certificate of successful completion with awarded contact hours will be mailed to each participant within 2 weeks of receipt of the **Registration Form, Post Test and Program Evaluation**.

Guidelines for Intravenous Prolastin-C Liquid Infusion

PURPOSE:
To provide specific guidelines for the intravenous administration of Prolastin-C® Liquid (Alpha$_1$-Proteinase Inhibitor Human).

INDICATION:
Intended to treat the unopposed protease activity, which causes pulmonary emphysema and necrotizing panniculitis in individuals with severe Alpha-1 Antitrypsin Deficiency. Not indicated for the treatment of liver disease due to Alpha-1 Antitrypsin Deficiency.

SELECTION CRITERIA:
Medical Criteria:

- Patient should be under the care of a licensed physician familiar with the use of Prolastin-C Liquid, including indications, dosing, method of administration, and potential adverse reactions.
- Consideration should be given to administering initial infusion in a medically supervised setting, however there is no requirement for this stated in the packet insert from Grifols Biotherapeutics, epinephrine injection/EpiPen and emergency equipment available.
- Establish home care services with a licensed home health agency/home infusion company with qualified IV nurses trained in the procedure. Some insurers do not cover home infusion services.

Additional considerations for home infusion:

- Patients who are motivated and physically able may be taught to self-administer Prolastin-C Liquid. Others may be infused by a significant other who has been properly trained in the procedure.
- The patient should be capable and feel comfortable with performing self-care safely after sufficient training and education. If not, the primary care giver must meet those requirements.
- It is important to have second responsible individual in the home during infusions.
- The home must have electricity, working phone and safe environment.
- A contingency plan should be in place for untoward events occurring before, during, and after the infusion.

HOW SUPPLIED:

- Trade name “Prolastin-C Liquid” manufactured by Grifols Biotherapeutics.
- Supplied in single use vials with the functional activity in milligrams stated on the label of each vial. Vial size is supplied in 1000 mg approximate activity. Milligrams per vial vary from lot to lot produced. Carefully read the prescription label and the milligrams contained in each vial since a given prescribed dose may require the pooling of varying numbers of vials per lot, depending on the milligrams per vial. It is usual practice, if a partial vial is calculated to complete the dose, to use the entire vial. Please contact the pharmacist if you have any questions or concerns regarding dosing or calculations.
- The approximate 1000 mg vials contain 20ml of solution for injection.

DOSE, ROUTE, and ADMINISTRATION:

- The recommended dose is 60 mg/kg body weight IV once weekly.
- Given by intermittent intravenous infusion.
- Recommended administration rate is 0.08 ml/kg/min, but this should be adjusted according to patient’s clinical condition and tolerance. Do not exceed rate of 0.08ml/kg/min.

- All connections for IV tubing's etc. for home infusion shall be leur lock
- Infusion can be administered via gravity infusion or IV pump. If the patient or caregiver administers without a nurse present, they will be taught to accurately regulate the IV rate.
- If the patient's prescription is for infusion other than weekly, it is recommended that weekly amounts are prepared and pooled, utilizing one set up of equipment per bag. This means that multiple bags may be administered.

DOCTOR'S ORDERS:
Will include:
- Dosage calculated according to patient's weight
- Route of administration
- Frequency of infusions
- Any premedications
- Laboratory monitoring parameters and schedule if indicated
- Standing order for use of epinephrine injection/EpiPen 0.3 mg IM prn anaphylaxis

CONTRAINDICATIONS
Do not use in patients with known selective severe IgA deficiency with antibody to IgA, due to the increased risk of severe hypersensitivity reactions, including anaphylaxis.

SIDE EFFECTS/POTENTIAL COMPLICATIONS
Side effects are reported to be mild and generally rare; they include:
- Chills
- Malaise
- Headache
- Rash
- Hot Flash
- Prurtis
- Anaphylaxis (rare) is a risk with any plasma product

NURSING IMPLICATIONS
- Assess needs of patient and/or primary caregiver: physical, psychosocial, & cognitive.
- Assess the patient's understanding of Prolastin-C Liquid therapy, potential side effects and risks of blood borne pathogen transmission.
- Educate the patient/caregiver against smoking, and exposure to second-hand smoke as smoke inactivates the drug and serves to further promote lung destruction.

- **Prolastin-C Liquid must be administered with inline filter of 5 to 15 micron.**
- **Administer within 3 hours after pooling. Keep Prolastin-C Liquid refrigerated at 2-8° C (36-46°F). Prolastin-C Liquid can be stored at room temp not exceeding 25 °C (77°F) for up to one month, after which the product must be used or immediately discarded.**

- Instruct patient and/or caregiver on proper storage of drug and supplies, therapy requirements, and procedures: Universal Precautions, aseptic technique, catheter care, connect, disconnect, medication preparation/handling and disposal of used supplies.
- Instruct patient in self-monitoring: including but not limited to: temperature, reporting of side effects, weight changes, over all status, including any changes in activity or exercise tolerance.
- Instruct patient to report significant changes to healthcare provider.
- If patient self-administers, plan home care follow-up with appropriate medical care team members at regular intervals. Establish a known and reliable contingency plan.
- Provide follow-up communication (verbal or written) including lab results, changes in condition to the physician, pharmacist, and other health care team members.
- Document all assessments, instruction sessions, home visits and follow-up plan.

- For home patient visits with RN performing the administration, documentation in the nursing notes should include:
    - Total Prolastin-C Liquid given milligrams and milliliters, Lot Numbers, concentration in vial, Exp. date
    - Route of infusion
    - Length of infusion
    - Baseline cardiopulmonary assessment (including breath sounds)
    - Baseline vital signs
    - Weight
    - Changes in condition, activity or exercise tolerance, sputum production, cough
    - Any patient complaints
    - Patient tolerance of infusion
    - Any adverse reaction and intervention

- Vital signs are assessed at baseline and at the end of infusion, as well as prn for any patient complaint during the infusion. Temperature is checked at baseline and end of infusion, or prn with patient complaint.
- Document and report any unusual or severe side effects to **Grifols Biotherapeutics at: 1-800-520-2807.**

PHARMACY IMPLICATIONS:
- Discuss procedure for obtaining prescriptions, renewals, and changes in therapy with the physician. Prolastin Direct must have accurate and signed prescription on file.
- Determine prescription appropriateness against the defined treatment goals and patient diagnosis.
- Document in the clinical record all communication with the physician, laboratories, home health nurse, and others involved in the patient's care.
- Discuss follow-up communication requirements (verbal or written) for the physician or other health care team members.

- **Prolastin-C Liquid must be administered with inline filter of 5 10 15 micron.**
- **Administer within 3 hours after pooling. Keep Prolastin-C Liquid refrigerated at 2-8° C (36-46°F). Prolastin-C Liquid can be stored at room temp not exceeding 25 °C (77°F) for up to one month, after which the product must be used or immediately discarded.**
- **The manufacturer recommends administering the solution within 3 hours of pooling.**

MISCELLANEOUS:.
- IV bag and IV tubing can be disposed in the household trash.
- Empty vials of Prolastin-C Liquid are to be disposed of in household trash. The only exception to this information is if your IV tubing has visible blood back up, at any time during the infusion then it should be considered biohazardous and discarded in the biohazard container. You should dispose of needles and other hazardous materials in the biohazard container.

References:
Physician's Desk Reference, 73rd Edition, 2017
Nursing IV Drug Handbook, Lippincott Williams & Wilkers, 2019
Grifols Biotherapeutics, Prolastin-C Liquid, Drug Package Insert August 2018

Instructions For
Prolastin®-C Liquid Home Mixing

Prolastin-C Liquid per patient prescription	18-gauge Needle(s)
Alcohol wipes	IV catheter/insertion supplies
Non-sterile gloves	15-micron filter IV administration tubing with
Empty IV bag(s)	Leur lock
Needle Free valve(s)	Tape
30ml Syringe(s) Luer lock tip	Bio waste container

Prolastin-C Liquid should be stored refrigerated at 2-8°C (36-46°F) for the period indicated by the expiration date on its label. Since refrigerated, remove Prolastin-C Liquid from the refrigerator prior to infusion and allow it to warm to room temperature. Prolastin-C Liquid may be stored at room temperature not to exceed 25°C (77°F) for up to one month, after which the product must be used or immediately discarded. Check lot numbers, mg of functional activity per vial and expiration dates and remove the correct number of boxes to prepare the prescribed dose.

Maintain strict asepsis while performing all preparation activities!

Pooling

Step 1. Identify and clean a designated work area. Gather supplies needed.

Step 2. Wash your hands.

Step 3. Obtain IV access prior to pooling medication in IV bag. Medication is only stable for 3 hours after it has been pooled, if not used within 3 hours Prolastin-C Liquid must be discarded.

Step 4. Remove the appropriate number of vials of Prolastin-C Liquid from their boxes.

Inspect the solution for particulate matter and discoloration prior to pooling. The product may contain a few protein particles. The solution is clear, colorless or pale yellow or pale green. Do not use if product is discolored or cloudy. Notify the pharmacy, as they want to retrieve that vial.

Step 5. Don non-sterile gloves.

Step 6. Remove the protective lids from the Prolastin-C Liquid vials, and aseptically wipe each rubber seal with an alcohol pad, using one pad for each vial, and allow to dry.

Step 7. Then locate the needle free valve and 18-gauge needle. Attach the needle free valve and 18-gauge needle.

Step 8. Wipe the rubber injection port of the empty IV bag with an alcohol pad and allow to dry. Remove the protective cap from the 18-gauge needle and insert the needle with the attached valve into the rubber injection port of the empty IV bag and secure the connection with tape.

Step 9. Remove the protective cap of the vented vial access spike. While holding the vial of the Prolastin-C Liquid firmly on the table, insert the spike into the center of the rubber seal.

Step 10. Once the vented access spike is inserted into the vial, then attach the sterile 30ml syringe to the vented access spike.

Step 11. Invert the bottle of Prolastin-C Liquid and withdraw the contents into the syringe.

It is essential to maintain aseptic technique. Remember to only handle the flanges of the syringe and spike, as they will be used repeatedly, proper handling of the syringe and vented vial access spike to avoid contamination is imperative.

Remember to wipe the needle free valve with alcohol and allow it to dry, each time before injecting into the bag.

Step 12. Once the contents have been drawn into the syringe, unscrew the syringe from the spike and attach the 30-ml syringe directly to the needle free valve at the injection port of the IV bag. Slowly inject the Prolastin-C Liquid into the IV bag to prevent foaming.

Step 13. Reattach the 30-ml syringe to the spike and remove the spike from the empty vial. Now insert it into the next vial, while maintaining aseptic technique.

Step 14. Repeat these steps until all vials are pooled into the IV bag.

Step 15. After all the vials are pooled into the IV bag, remove the needle free valve and 18-gauge needle from the injection port and dispose of it in the bio waste container.

Step 16. Discard all empty Prolastin-C Liquid vials into the household trash or recycling container. Used needles must be discarded into the bio waste container.

Step 17. Prolastin-C Liquid is now ready for the infusion. Attach the 15-micron filter IV administration tubing and prime.

NOTE: At the end of the infusion, all IV bags and administration tubing can be disposed of into the household trash unless contaminated with visible blood at any time during the infusion. If blood has been observed, the IV bags, syringes and IV administration tubing must be disposed of into the bio waste container.

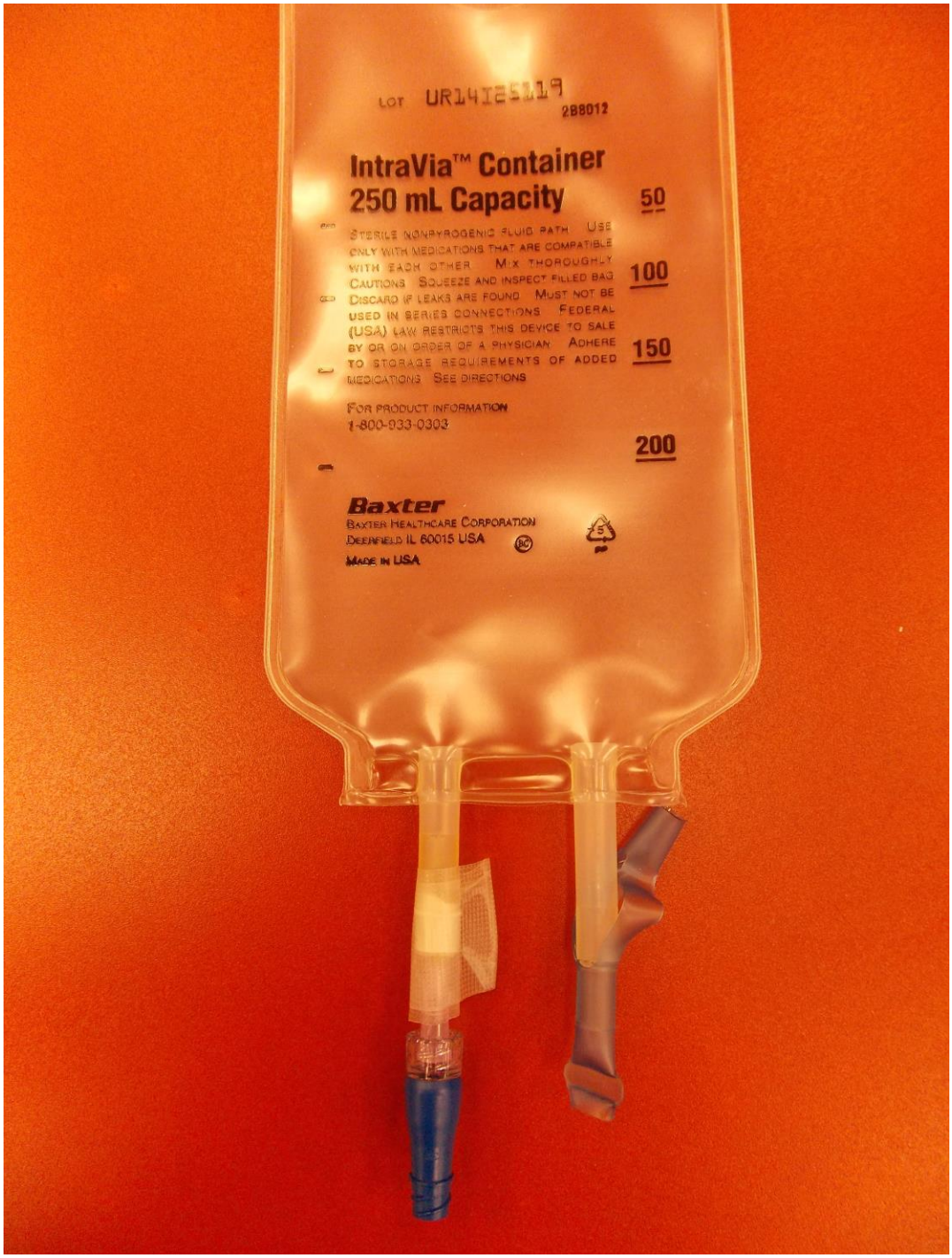
Needle Free Valve

Similar item available from various manufacturers

Vented Vial Access Spike

Similar item available from various manufacturers

Bag with Needle Free Valve



Infusion Calculations

RECOMMENDED DOSE: 60 mg/kg IV weekly

*****Prolastin-C Liquid is available in approximate 1000mg vials with 20 ml of solution for injection.**

*****Milligrams contained in each vial vary from lot to lot produced*****

Carefully read the prescription label and the mg amounts contained in each vial. Doses given are equal to or slightly greater than the calculated dose due to the varying assays. No drug is withdrawn or wasted. The number of vials pooled to achieve the patient's dose will vary from lot to lot. One cannot safely assume that the same number of vials is pooled for each infusion prepared from subsequent lots.

Please contact the pharmacist if you have any questions or concerns regarding dosing or calculations.

DOSE CALCULATION:

Example: Individual weighing 155 lbs.

Step 1:

Convert lbs. to kg by dividing by 2.2

155 ÷ 2.2 = 70.5 kg

Multiply 60 mg (prescribed weekly dose) x 70.5 kg= 4230 mg

Dose is 4230 mg IV weekly

Step 2:

Determine how many vials of Prolastin-C Liquid will be needed to formulate the required dose.
For our example we will use a lot with 1078 mg per vial.

Divide the calculated dose of 4230 mg by the number of milligrams in a vial.

4230mg ÷ 1078mg = 3.9 vials

Remember that no drug is wasted: therefore, total vials to be infused will be 4.
Total mgs are 4312 mg and total fluid volume: 80 ml (4 x 20 ml)

RECOMMENDED INFUSION RATE: 0.08 ml/kg/min

RATE CALCULATION:

Using the same example of a 70.5 kg individual:

To calculate the patient's infusion rate using the recommended rate of 0.08 ml/kg/min you would do the following steps

Step 1. Multiply 0.08 ml x 70.5 kg = 5.64 ml/min

Step 2. Convert ml/min to ml/hr by multiplying by 60 (60 min/hour)
5.64 ml x 60 min = 338 ml/hr

Step 3. Calculate the drop rate using the formula as shown

$$\frac{\text{Drops (gtt) of IV set}}{60 \text{ (min in hour)}} \times \text{total hourly volume} = \text{gtts/minute}$$

In this example, the calculation would look like this:

$$\frac{\text{10 gtts}^{**}}{\text{60 min/hr}} \times 338 = 56 \text{ drops/min}$$

**** Remember to check the drop factor of your specific IV tubing as Drop factor rates vary**

Step 4. Divide 56 by 4 as there are four 15-second periods in a minute

56 ÷ 4 = 14 drops

Step 5: Count 14 drops/15 seconds to regulate the rate