

# Medications for Alpha-1 Lung Disease

Skinny Little Reference Guide™



## INTRODUCTION

In general, the medicines used to help Alphas with their breathing difficulties are the same as those prescribed for individuals with asthma, chronic bronchitis, emphysema (including emphysema associated with Alpha-1 Antitrypsin Deficiency), COPD, and bronchiectasis. Some of these medications can be taken as pills or capsules, and some are inhaled as a mist or a powder. Occasionally, during episodes of severe respiratory distress, some medications may be given by injection.

To help you learn more about your medications, the following pages include descriptions of the most common medicines prescribed for individuals with Alpha-1 lung disease. One specific type of medication, augmentation therapy, is not included in this brochure. Because of its important place in the treatment of Alpha-1 lung disease, information about augmentation therapy is presented in a separate brochure titled "Augmentation Therapy: The Specific Treatment for Alpha-1 Lung Disease." Additionally, you can find a detailed discussion of augmentation therapy in the Big Fat Reference Guide™ at [www.alphanet.org](http://www.alphanet.org)



**CROSS REFERENCE:** For detailed information about augmentation therapy see the brochure "Augmentation Therapy: The Specific Treatment for Alpha-1 Lung Disease," or the section on "Augmentation Therapy" in the Big Fat Reference Guide™ at [www.alphanet.org](http://www.alphanet.org)

Because information about medications can be confusing we have attempted to simplify the information by listing similar types of medications according to how these medications work. These categories are referred to as "drug classifications." In this way, we can describe the characteristics and common side effects for many different kinds of drugs based on their respective drug classifications. We also have included some trade names and generic names of drugs that fall into each of these classifications.

However, keep in mind new drugs are constantly becoming available. If your doctor has prescribed a medication for you that you cannot find in these lists, ask your healthcare provider to tell you in which "drug classification" your medication belongs.

Each of these summaries includes a list of side effects that may be associated with a specific classification of drugs. The purpose for listing all of these potential side effects is to alert you to the possibility of these occurrences, but it does not mean that you can expect to experience all, or any, of these effects. Some critical side effects are rarely observed, but they are listed so you can recognize them should they occur.

Be aware that no list of side effects can include all the problems that an individual might experience with a particular medication. Therefore, you should always discuss any side effects or unusual symptoms you experience with your healthcare provider.

## HOW THESE MEDICATIONS WORK

The various medications commonly prescribed for lung disease work in many different ways, including:

- Decreasing inflammation and mucus production
- Preventing and/or decreasing pulmonary exacerbations
- Opening the airways so mucus can be removed
- Preventing and relieving trapped air in the lungs
- Relieving constriction of the bronchial tubes, allowing for better movement of air in and out of the lungs
- Fighting the infections that can accompany an exacerbation

Medications that are taken as pills, capsules, liquids, or by injection are said to have "systemic effects," meaning these drugs enter the bloodstream and can affect the body as a whole. Inhaled medications, on the other hand, enter the lungs directly and, therefore, their effect on other parts of the body tends to be minimized. Some medications are prescribed to be taken on a daily basis and others should be taken only when the need arises. To make sure you understand how and when you should take your medications, review your list of medications with your healthcare provider.

It is often helpful to prepare a list of your medications. Your medication list should include:

- All the medications you are taking on a routine basis including over-the-counter and alternative-type medications
- Medications that you take only when you develop certain symptoms
- The order and time of the day to take medications
- The methods for administering medications (e.g., pills, inhalers)
- A plan for adjusting dose levels or frequency of administration during times of severe respiratory distress or exacerbations



**CROSS REFERENCE:** To make a list of your medications see the "My Medications" worksheet located in the Big Fat Reference Guide™ at [www.alphanet.org](http://www.alphanet.org)

For quick reference complete the Medical Information Worksheet on the last page of this brochure, cut it out and keep it in your wallet.

Now, let's look at each drug classification with its description of the benefits and possible side effects of the common medicines used to treat lung disease.

## BETA-AGONISTS

Beta-agonists are used to relax the muscles that surround the bronchial airways when they become constricted. Drugs in this classification are derived from adrenaline, a natural hormone associated with a rapid surge in energy, such as when you are frightened, startled, or excited. They can be administered by an inhaler or nebulizer, orally (by a pill or capsule), by a "subcutaneous" injection under the skin, or by intravenous infusion (IV). Inhaling the drug directly into the airways of the lung is the preferred way these drugs are administered, because this reduces the potential for systemic side effects. Possible side effects from beta-agonists include: shaking, trembling, rapid heart rate, dizziness, and anxiety.

### USING SHORT-, MEDIUM-, AND LONG-ACTING BETA-AGONISTS

Initially beta-agonists, such as adrenaline, were found to be quite effective and were often prescribed. However, because their benefit was short-acting in nature (effective duration of 1- 2 hours), scientists have developed beta-agonists that are longer acting, thereby improving this class of drugs. Medications with short duration are used in extreme emergency situations, such as severe allergic reactions. If it is necessary to use this type of medication, you must be seen in an emergency room for evaluation as soon as possible.

The medium-acting bronchodilators can be used on a regular basis, or as "rescue" drugs. Rescue drugs, such as albuterol, pirbuterol, metaproterenol, or terbutaline, have a rapid onset of symptom relief. You may require a rescue bronchodilator from time to time, for example, during periods of physical exertion, or if you come into contact with high levels of respiratory irritants or pollutants. Medium-acting bronchodilators start working within 5 minutes, and their effects can last 4-6 hours.

The long-acting bronchodilators last for 12 hours, but they require up to 30 minutes after dosing before their full effects are felt. Long-acting bronchodilators should never be used as a rescue medication. Using long-acting bronchodilators on a routine basis will help maintain an open airway throughout the day and night. Beta agonists with even longer durations of action are currently under development.

#### COMMONLY USED BETA-AGONISTS

U.S. TRADE NAME	GENERIC NAME	INH* (MCG)**	NEBULIZER (MG)	ORAL (MG)	DURATION OF ACTION (HRS)
<b>SHORT-ACTING</b>					
	adrenaline or epinephrine***	N/A	N/A	N/A	1-4
<b>MEDIUM-ACTING</b>					
Alupent	metaproterenol	650	10-15	-	4-6
Ventolin, Proventil, Volmax, Repetabs, Combivent	albuterol	100-200	2.5-5.0	4	3-6
Maxair	pirbuterol	250-500	5-10	-	3-6
Bricanyl, Breathine	terbutaline		2.5-5.0	-	3-6
<b>LONG-ACTING</b>					
Serevent, Advair	salmeterol	50-100	0.02	-	12+
Foradil, Perforomist, Symbicort	formoterol	12-24	0.02	-	12+

\* INH = inhaled medicine given by metered dose inhaler (MDI) or dry powder inhaler (DPI)

\*\* Important to note that mg's are 1,000 times greater than mcg's

\*\*\* Given by subcutaneous injection, also known as EPI

## ANTICHOLINERGICS

Like the beta-agonists, anticholinergics also relax the muscles of the airways, but they accomplish this in a different way. Anticholinergics block the nerve impulses that normally stimulate the airways to constrict and reduce the amount of secretions in the airways. These drugs are available as inhalers, as liquids to add to a nebulizer, and as nasal sprays. Recent clinical studies in COPD have provided information suggesting anticholinergics can be used as a "first line of defense" to prevent exacerbations.

You should be aware that anticholinergics may have an additive effect when taken in combination with beta-agonists. An "additive effect" means you can take these two classes of drugs in combination and get a more powerful effect than if you were to take either drug separately.

Anticholinergics may cause dryness of the mouth, cough, nervousness, headache, and in older men in particular, difficulty with urination. Although anticholinergics also can dilate the pupils of your eyes, leading to increased pressure that may result in worsening of certain forms of glaucoma, this is not generally a concern for the inhaled form of the drug (unless it is sprayed, or rubbed, directly into the eyes).

#### COMMONLY USED ANTICHOLINERGICS

U.S. TRADE NAME	GENERIC NAME	INH* (MCG)**	NEBULIZER (MG)	ORAL (MG)	DURATION OF ACTION (HRS)
<b>MEDIUM-ACTING</b>					
Atrovent, Combivent	ipratropium	40-80	0.25-0.5	-	6-8
<b>LONG-ACTING</b>					
Spiriva	tiotropium	22.5	-	-	24

\* INH = inhaled medicine given by metered dose inhaler (MDI) or dry powder inhaler (DPI)

\*\* Important to note that mg's are 1,000 times greater than mcg's

## INHALED STEROIDS

Inhaled steroids are used to decrease inflammation and exacerbations, and are effective in treating COPD and asthma-like symptoms. Steroids taken orally or given by injection can be life-saving, but also can produce serious side effects. To reduce these undesired effects, new forms of steroids were developed that can be inhaled directly into the lungs. These medications are almost always administered by an inhaler. The use of steroids by inhalation has greatly improved the safety of this class of medication; however, they may be ineffective for very severe cases. This is due, in part, to the fact that inhaled medications cannot get to the sites in the lung most affected when the airways are blocked by mucus, sputum, infected material, and airway closure.

It also should be kept in mind that the inhaled form of steroids can still be absorbed into the bloodstream (especially with the newer, more potent brands taken at high doses) and exert an effect on the whole body. Therefore, if your doctor prescribes the temporary use of an oral steroid during an exacerbation, you should verify whether or not you should also continue taking an inhaled steroid medication during this same time period. If your doctor advises you to temporarily discontinue an inhaled steroid medication, make sure you also ask when the inhaled steroid can be restarted.



**KEY LEARNING:** If your doctor prescribes the temporary use of an oral steroid during an exacerbation, you should verify whether or not you should also continue taking an inhaled steroid medication during this same time period.

Although many of the severe steroid side effects can be eliminated or greatly reduced by the use of inhalers, one side effect that is actually increased is the development of thrush, an oral yeast infection. This is characterized by a severe sore throat and extreme redness of the tongue, with white patches in the mouth and/or back of the throat, along with the possibility of a metallic-like taste (see page 16). Swallowing may be quite painful as well.

If this should occur, it often is necessary to temporarily reduce or eliminate the use of the steroid inhaler. Occasionally, these undesirable side effects may persist, in which case it may be necessary to permanently discontinue the use of steroid inhalers.

Antifungal medications can be prescribed for the treatment of thrush; however, you may be able to prevent its development by using a "spacer" (see "Using Spacers with Inhalers" on page 9). Using a spacer also will help prevent the absorption of the steroid medication into the bloodstream, which could reduce the incidence of other side effects associated with the use of steroids. You can further reduce the incidence of other undesired side effects by rinsing your mouth after using a steroid inhaler. It also can be helpful to use your inhaler just before you brush your teeth in the morning and evening.

### COMMON INHALED STEROIDS

TRADE NAME	GENERIC NAME	FORM*
Aerobid	flunisolide	INH
Beclovent, Vanceril, Q-var	beclomethasone	INH
Flovent, Advair**	fluticasone	INH
Azmacort	triamcinolone	INH
Pulmicort, Symbicort**	budesonide	INH

\* INH = inhaled medicine given by metered dose inhaler (MDI) or dry powder inhaler (DPI)

\*\* a combination drug (also see beta-agonist classification)

## PROPER USE OF INHALERS

Some medications are available in an inhaled form administered by a device called an inhaler. Inhalers are more formally known as metered dose inhalers, or MDIs, because they deliver a fixed, specific dose with each activation. Due to the worldwide ban on the use of fluorocarbons, virtually all MDI's are being reformulated for use as non-fluorocarbon or non-propellant inhalers. Fluorocarbons present an environmental hazard as they are known to contribute to the destruction of the ozone layer.

## SPRAY-TYPE METERED DOSE INHALERS (MDIs)

The technique you use with metered dose inhalers (MDIs) can affect how much of the drug reaches the airways of the lung. Your doctor or healthcare provider can advise you on which technique is best for you, depending on the type of medication prescribed. For spray-type inhalers, without a spacer, do not place the inhaler directly in the mouth, unless specifically instructed to do so by your healthcare provider, because this can cause a large amount of the drug to be deposited on the tongue and in the back of the throat (instead of in the lungs where you need it).

This type of metered dose inhaler consists of a pressurized canister of medication in a plastic case with a mouthpiece. Pressing the MDI releases a mist of medication. When inhaled correctly, the medication has a better chance to reach the small airways and increases the medication's effectiveness.

The following steps outline the best method to use an MDI:

1. Remove the cap from the inhaler
2. Hold the inhaler with the mouthpiece at the bottom
3. Shake the inhaler — this mixes the medication properly
4. Hold the mouthpiece 1-1/2 to 2 inches (2 - 3 finger widths) in front of your mouth. This improves medication delivery by slowing mist delivery and only allowing small particles to be inhaled. (Figure A); Note that some products are best used by placing the inhaler in your mouth (Figure B), and you should check product instructions for your type
5. Gently breathe out
6. Tilt your head back slightly and open your mouth wide
7. Press the inhaler and at the same time begin a slow, deep breath
8. Continue to breathe in slowly and deeply over 3 - 5 seconds; breathing slowly delivers the medication deeply into the airways
9. Hold your breath for up to 10 seconds; this allows the medication time to deposit in the airways
10. Resume normal breathing
11. Repeat steps 3 - 10 when more than one puff is prescribed

Note: MDIs should not be left in conditions of extreme heat or cold as this can impair their function.



**KEY LEARNING:** It is important to keep your inhaler clean. Some products suggest using a wet Q-Tip to clean the opening. Follow the instructions in your product packaging and/or check with your healthcare provider for more specifics.

## USING SPACERS WITH INHALERS

Another method for improving the delivery of inhaled medication involves the use of a device called a "spacer" (Figure C). Delivering medications to the lung is a combination of drug formulation, delivery system properties, the patient's inhalation technique, and the geometry of the airways. Fine-particle, low-velocity aerosols carry drugs more deeply into the lungs. Most of the side effects incurred with the use of inhalers are because of large particles of medication becoming trapped in the mouth.

The reasons a spacer is recommended are:

- Larger particles of the drug are trapped in the spacer itself (not in your mouth)
- The fine mist is inhaled deeper into the small airways
- The risk for potential side effects is decreased
- You do not have to coordinate the timing of the activation of an MDI with your breathing

Several types of spacers are available, and you can select the kind that works best for you. Each spacer should come with a specific set of instructions regarding its use and cleaning. Keeping the spacer clean and dry is important. You should be aware that water left standing in a spacer could allow for the growth of bacteria that could then be inhaled into your lungs. It is important to allow the spacer to properly dry after each cleaning.

## DRY POWDER INHALERS (DPIs)

Dry powder inhalers are becoming more common, in part because they do not use the CFC propellant found in spray-type MDIs. CFCs damage the ozone in the atmosphere and will be phased out of MDIs in the next few years. Dry powder inhalers are as effective as MDIs, and in fact, may prove to be slightly more effective.

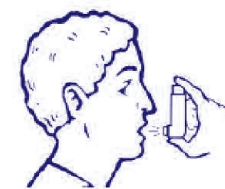


Figure A

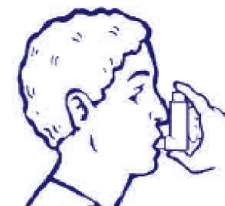


Figure B

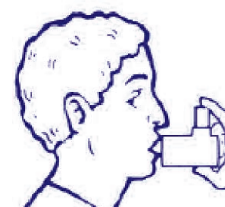


Figure C





This device is similar to an MDI, but the drug is in powder form. Both are handheld devices that deliver a precisely measured dose of medicine into the lungs. The advantage of using a dry powder inhaler is that it is breath-activated, so you don't have to coordinate activating the inhaler while at the same time inhaling the medication. Instead, you simply breathe in quickly to activate the flow of medication. In this way, the breath-activated discharge of medicine is always coordinated with your inhalation effort. There are other important differences, so make sure that you always know the proper technique for the method you are using.

There are many types of dry powder inhalers available. Each of them has a different operating method. Some have to be loaded each time they are used (Inhalator, Spinhaler, Rotohaler). Others have disks with a set number of doses (4 or 8), while still others have as many as 200 doses stored in the device (Turbuler, Accuhaler). To get the full benefit from these DPIs, you need to make sure you hold your breath for about 10 seconds after taking the medication.

Another example of a specific type of dry powder inhaler is a Diskhaler. For proper use of this device, follow the instructions included with the Diskhaler package.

There also are a number of disadvantages associated with the use of DPIs. If you exhale directly toward the device, the powder can be blown out. Also, much of the powder ends up in the mouth, which can cause unwanted side effects. As with MDIs, it is recommended you rinse your mouth after administering the drug.

## NEBULIZERS

A nebulizer is another device that delivers medications such as beta-agonists and anticholinergics in a fine spray or mist. MDIs, dry powder inhalers, and nebulizers are all important therapy options; however, these delivery systems are not equally suitable for all patients. In general, the physician's first choice of aerosol delivery for beta-agonists, anticholinergics, and steroids is the MDI or DPI; however, nebulizers remain useful for several reasons. First, some drugs for inhalation are available only in solution form. Second, some patients cannot master the correct use of MDIs or dry powder inhalers. Third, some patients prefer the nebulizer to other aerosol-generating devices.

The physiologic benefits of MDIs and nebulizers are virtually equivalent, and the choice of device is often based on the healthcare provider or patient preference. Sometimes, medical insurance reimbursement policies, rather than superiority of one approach over the other, may dictate which system you use. To determine which system may be best for you, discuss the risks and benefits on the following page with your healthcare provider.

## ADVANTAGES AND DISADVANTAGES OF NEBULIZERS

ADVANTAGES	DISADVANTAGES
It often is recommended for people who cannot use other inhaler devices.	The side effects of anxiety and increased heart rate can be magnified because of the increased dosage of the medication administered.
The medication is delivered either through a mouthpiece or with a facemask that is not dependent upon the person's breathing technique. Coordination of breathing and medication delivery is NOT necessary. Thus, if a patient cannot master the technique required of an MDI or has increased shortness of breath, a nebulizer may be the device of choice.	Routine use of nebulized medications, in combination with a long-acting beta-agonist inhaler, increases the risk of severe side effects. Anxiety and increased heart rate, along with sensations of increased shortness of breath, may indicate you are getting too much medication.
Nebulizers deliver a higher dose of medication – up to 30 times the amount delivered by a metered dose inhaler.	A nebulizer treatment is time-consuming and requires equipment to administer the medication. It also may limit access when you are away from home, resulting in missed doses. Equipment must be cleaned after every use.
It may provide relief during severe respiratory attacks, because the medication is delivered continuously for 10 to 15 minutes.	Infections: strict cleaning techniques must be done on a routine basis; otherwise infections may occur due to waterborne bacteria, such as pseudomonas. Leaving water standing in the nebulizer when it is not in use allows bacteria to grow.



## STERIODS (CORTICOSTEROIDS)

The steroid medications prescribed for lung disease should not be confused with the anabolic steroids that are sometimes used by athletes to increase their muscle mass. Corticosteroids are the most potent medications available for the treatment of severe obstructive lung disease and a variety of other inflammatory conditions. Although the effectiveness of these drugs can be life-saving in many cases, the potential for severe side effects often goes hand-in-hand with this class of drug, especially if given in high doses or for a long period of time.



**BURNING ISSUE:** Corticosteroids are the most potent medications available for the treatment of severe obstructive lung disease and a variety of other inflammatory conditions. Although the effectiveness of these drugs can be life-saving in many cases, the potential for severe side effects often goes hand-in-hand.

Steroids are naturally occurring hormones produced primarily by the adrenal glands, which are located directly above the kidneys. These hormones play an important role in maintaining metabolism and a number of other complex interactions within the body as a whole. Since steroids have such wide-ranging effects in addition to their anti-inflammatory properties, their use must be monitored closely and they should be used with caution. Steroids may be taken systemically (by mouth or by injection) or by inhalation (through an MDI or nebulizer). Some of the differences between how these medications work with each of these administration methods are described below.

The mechanisms by which steroids help asthma, emphysema/COPD, and other chronic lung conditions is not fully understood and remain under intense investigation.

### SYSTEMIC STEROIDS (ORAL MEDICATIONS OR INJECTIONS)

When a medication is given "systemically," it means that the drug enters the bloodstream and exerts its effect throughout the entire body. This effect is achieved when the medication is taken orally, as a pill or a liquid, or when it is given by injection into the muscle or directly into a vein. Systemic corticosteroids are prescribed for individuals with extremely severe Alpha-1 lung disease, or to Alphas when they experience severe exacerbations of their lung disease.

Among the most dramatic effects of steroids is the suppression of inflammation. This can be a good effect if you have a disease characterized by excessive inflammation, such as asthma or arthritis. But suppression of inflammation can be a problem if the inflammation is serving a useful purpose. For instance, individuals on systemic steroids can develop life-threatening bacterial infections without being aware of it because the steroids are suppressing the body's inflammatory response to the infection. Thus, individuals receiving steroids who develop severe infections may not experience fever, show redness, swelling, or have pain at the site of an infection. For these reasons, long-term systemic steroid use is generally avoided in all but the most extreme cases of disease.



**KEY LEARNING:** Long-term systemic steroid use is generally avoided in all but the most extreme cases of disease.

Caution also needs to be taken when steroid medications are discontinued. This is because when individuals receive supplemental steroids systemically for more than four to five days, the adrenal glands can start to shut down their own steroid production, and the pituitary gland at the base of the brain can stop sending signals to the adrenals to make steroids. In these cases, if steroid medication was to be suddenly discontinued, the adrenal glands would not be able to immediately resume adequate steroid production. The lack of adequate levels of steroids could then lead to the development of life-threatening situations. Therefore, when supplemental steroids are no longer required, the dose levels are gradually tapered over a period of time. By tapering the dose of steroid medication, the adrenal glands have a chance to recover their normal production, which reduces the risk of developing adverse effects.



**KEY LEARNING:** When supplemental steroids are no longer required, the dose levels are gradually tapered over a period of time.

If you have been gradually weaned from steroids, you still may require supplemental steroidal medication for exacerbations or during stressful situations, such as if you were to suffer a serious injury or require surgery. In these events, be sure to inform your physician if you have been weaned from steroid use within the past year. It also is recommended that individuals who are receiving chronic systemic steroids, or who have been weaned from chronic systemic steroid use within the previous year, should wear a medical alert bracelet or necklace with the terms "steroids" or "chronic steroids" inscribed on it.





**REMEMBER:** If you have been gradually weaned from steroids, you still may require supplemental steroidal medication for exacerbations or during stressful situations, such as if you were to suffer a serious injury or require surgery.

## SYSTEMIC STEROID MEDICINES

TRADE NAME (EXAMPLES)	GENERIC NAME	FORM*
Solu-Medrol Depo-Medrol Medrol	methylprednisolone	IV, IM IV, IM Oral
Solu-Cortef	hydrocortisone	IV
Celestone	betamethasone	IM
Multiple	prednisone	Oral
Multiple	prednisolone	Oral

- IV = intravenous infusion
- IM = intramuscular injection
- Oral forms may be pills, capsules or liquids

## ROUTINE STEROID USE AND SIDE EFFECTS

The use of routine steroid pills or frequent steroid bursts can cause a number of side effects. Steroid side effects usually occur after long-term use with high doses of steroid pills. In addition, high doses of inhaled steroids can cause some of these same side effects.

## SIDE EFFECTS THAT MAY BE ASSOCIATED WITH HIGH-DOSE STEROID PILLS INCLUDE:

SIDE EFFECTS	PREVENTION OF SIDE EFFECTS
Endocrine (hormones): <ul style="list-style-type: none"> <li>• Suppression of the adrenal and/or pituitary glands</li> <li>• Delayed sexual development</li> <li>• Changes in menstrual cycle</li> <li>• Increase and change in fat placement, causing fullness the face, "widow's hump," and weight gain</li> <li>• Increased blood sugar (diabetes)</li> <li>• Emotional changes such as moodiness, depression, euphoria or hallucinations</li> </ul>	Endocrine (hormones): <ul style="list-style-type: none"> <li>• Your healthcare provider may prescribe your steroid pills at specific times; make sure you take them as prescribed and do not stop them suddenly</li> <li>• If you have taken oral steroids, talk with your healthcare provider about obtaining a medical alert bracelet</li> <li>• Talk with your healthcare provider if you are having moodiness or depression that doesn't seem to get better</li> </ul>
Fluid and electrolytes <ul style="list-style-type: none"> <li>• Salt and water retention</li> <li>• High blood pressure (hypertension)</li> <li>• Loss of potassium</li> </ul>	Fluid and electrolytes <ul style="list-style-type: none"> <li>• Limit the amount of salt and foods that are high in sodium to prevent fluid retention and swelling; condiments and processed foods tend to be high in sodium</li> <li>• Add foods that are high in potassium to your diet</li> </ul>
Eyes <ul style="list-style-type: none"> <li>• Increased pressure in the eye (glaucoma)</li> <li>• Clouding of vision in one or both eyes (cataracts)</li> <li>• Blurred vision</li> </ul>	Eyes <ul style="list-style-type: none"> <li>• Visit the eye doctor (ophthalmologist) at least yearly; inform him or her that you take steroid pills routinely</li> </ul>
Skin <ul style="list-style-type: none"> <li>• Increase in body hair and acne</li> <li>• A tendency to bruise easily</li> <li>• Thinning of the skin and poor wound healing</li> </ul>	Skin <ul style="list-style-type: none"> <li>• Ask your healthcare provider about how acne can be treated</li> <li>• Keep the skin well moisturized</li> </ul>
Nutrition <ul style="list-style-type: none"> <li>• Increase in appetite</li> <li>• Irritation of stomach and esophagus with possible ulcer symptoms and rarely, bleeding</li> </ul>	Nutrition <ul style="list-style-type: none"> <li>• If you are eating more food, be sure you choose low-fat, low-sugar items to control calories – ask your healthcare provider or dietitian to help you with a specific diet plan</li> <li>• Eat a well-balanced diet that meets the Food Pyramid Guidelines</li> <li>• Take your steroid dose with food to decrease stomach irritation</li> </ul>

(CONTINUED)

SIDE EFFECTS	PREVENTION OF SIDE EFFECTS
Muscles <ul style="list-style-type: none"><li>• Muscle weakness or cramps</li></ul>	Muscles <ul style="list-style-type: none"><li>• Routine exercise may be recommended to prevent or decrease muscle weakness</li></ul>
Bones <ul style="list-style-type: none"><li>• Joint pain (especially as steroids are decreased)</li><li>• Thinning of bones (osteoporosis) may lead to fractures or compressions, especially of the backbone and the hip</li><li>• Loss of blood supply to bones (aseptic necrosis) may cause severe bone pain and require surgical correction</li></ul>	Bones <ul style="list-style-type: none"><li>• To prevent osteoporosis (loss of calcium in the bones), it is important to eat foods high in calcium, such as dairy products – if you need to control calories, low-fat dairy products may be used</li><li>• Your healthcare provider or dietician may recommend certain supplements, such as calcium, vitamin D, and a multi-vitamin</li><li>• Weight-bearing exercise also may be recommended by your healthcare provider</li><li>• Medication may be prescribed to improve osteoporosis</li></ul>
Immune System <ul style="list-style-type: none"><li>• General suppression of the immune system causes an increased risk to a variety of infections, for example, chicken pox</li></ul>	Immune System <ul style="list-style-type: none"><li>• Good hand-washing</li><li>• Avoid exposure to any infectious disease</li><li>• If you or your child is exposed to chicken pox or measles while receiving oral steroids or high-dose inhaled steroids, notify your healthcare provider immediately to determine if any special treatments needed</li></ul>

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WHAT ABOUT THRUSH?

Another common side effect associated with the use of inhaled steroids is thrush. Candidiasis of the mouth and throat, also known as "thrush" or oropharyngeal candidiasis (OPC), is a fungal infection that occurs when there is an overgrowth of fungus called Candida. Candida is normally found on the skin or mucous membranes. However, if the environment inside the mouth or throat becomes imbalanced, Candida can multiply. When this happens, symptoms of thrush appear.

**What are the symptoms of thrush?** Individuals with OPC infection usually have painful, white patches in the mouth. Symptoms of OPC in the esophagus may include pain and difficulty swallowing.

**How do I get thrush, and is it preventable?** Individuals at highest risk of OPC include persons receiving antibiotics, diabetics, smokers, and denture wearers. Candida (yeast) infection in the mouth is frequently caused by residue from metered dose inhalers (MDIs), particularly steroid inhalers. Use of a spacer device can help eliminate thrush. Another method to reduce the incidence of thrush is to rinse the mouth out with water and spit the water out immediately after an inhaler dose. Water is better than using a mouthwash, since it will help maintain the natural flora of the mouth and throat. Unsweetened yogurt with a capsule of acidophilus dissolved in it can help replace the natural bacteria and reduce the Candida overgrowth. Fruit can be added to the yogurt for better flavor. Acidophilus is available in health food stores and pharmacies. It can also help control diarrhea that accompanies antibiotic therapy.

**What is the treatment for thrush?** Prescription treatments, such as oral fluconazole or nystatin suspension, usually provide effective treatment for OPC.

THEOPHYLLINE AND AMINOPHYLLINE

These drugs have been used for over a century and are some of the earliest therapies discovered that help improve difficult breathing. Closely related to caffeine, theophylline and aminophylline belong to a classification of chemicals known as methyl-xanthines. One way in which these drugs work is by blocking the production of a particular enzyme, phosphodiesterase, which then promotes a series of cellular events that result in relaxation of the muscles of the lung's airways.

These drugs are not prescribed as frequently as they once were due to their potential to produce severe side effects. Individuals taking these medications may be required to obtain routine blood tests to measure the amount of theophylline accumulating in the blood, and ensure these levels do not become too high. Potential side effects include: nausea, vomiting, shakiness, and sleeplessness. If the theophylline blood level becomes too high, severe side effects may include seizure and death. Certain medications can dramatically increase Theophylline levels including erythromycin antibiotics, certain antihistamines, and Cipro and related antibiotics.

TRADE NAME (EXAMPLES)	NAME	FORM
Theo-Dur, UniDur Choleryl, Theo24 Slo-Bid, Slo-Phyllin	theophylline	Oral
	aminophylline	IV

\*Oral forms may include pills, capsules, or liquids

\*IV = intravenous infusions

## MAST CELL STABILIZERS

Mast cells are a specific type of cell found in connective tissue in the lungs. When these cells interact with certain types of antibodies they burst, releasing a variety of chemicals that can produce asthmatic symptoms. Mast cell stabilizers prevent the cells from bursting. In this way, this type of medicine minimizes the effects of these cells and produces an improvement in breathing. Although these drugs are in limited use, they are available as inhalers or a liquid that can be added to a nebulizer. This class of medication is prescribed to prevent the onset of breathing difficulties and is not effective for the treatment of exacerbations.

Although their benefit for COPD has not been proven, mast cell stabilizers may be indicated for individuals with reactive airways disease. To achieve effectiveness, these medications must be used on a continual basis, rather than just when breathing appears to be impaired. Side effects are generally minimal; however, the powdered form of these medications can occasionally produce asthmatic episodes in some individuals.

TRADE NAME (EXAMPLES)	GENERIC NAME	FORM*
Intal Tilade	cromolyn nedocromil	INH INH

\*INH = inhaled medicines may be given by metered dose inhalers (MDIs), dry power inhalers (DPIs) or nebulizers

## ANTI-LEUKOTRIENES

Leukotrienes are chemicals manufactured by the body that induce an inflammatory response. When the level of lung leukotrienes is heightened, airway constriction can occur. Anti-leukotrienes are a new class of medication available only in a pill form that block the effect of these chemicals and help open breathing passages. These medications are usually prescribed for individuals with asthma, and it is not yet clear whether or not these medications are beneficial in patients with COPD.

The incidence of side effects is reported to be low with these drugs, but may include headache, upset stomach, weakness, and flu-like symptoms. Of particular importance to Alpha-1 individuals, anti-leukotrienes may interfere with the normal function of the liver. Blood tests to monitor the level of certain liver enzymes can be done on a routine basis to determine whether or not these drugs are causing adverse effects in the liver. In addition, special precaution appears to be warranted in individuals who are being weaned from steroids, because anti-leukotrienes have been shown to have serious side effects in these patients.

TRADE NAME (EXAMPLES)	GENERIC NAME	FORM*
Singulair Accolate Zyflo	montelukast zafirlukast zileuton	oral oral oral

\*Oral forms = pills

## MUCOLYTICS

Some individuals with Alpha-1 COPD may experience breathing difficulties when they develop large amounts of thick, sticky mucus that seem to become trapped in the throat or the lungs. Mucolytics are used to thin and liquefy thick, sticky secretions in the lungs so they then can be more easily coughed up from the respiratory tract.

The most commonly used mucolytic is acetylcysteine (Mucomyst). The standard dose of acetylcysteine is 2 - 4 cc of the 10% or 20% solution given by nebulizer, mixed with a bronchodilator. Frequently, 2.5 mg albuterol is the bronchodilator of choice to mix with acetylcysteine. These medications are given 3 or 4 times daily, until the thickness of the secretions decreases to a manageable level. Acetylcysteine should be stored in a refrigerator after the package or container has been opened, and must be used within 96 hours.

In special cases, such as for cystic fibrosis or severe bronchiectasis due to Alpha-1, a special type of mucolytic called Pulmozyme or dornase alpha is sometimes prescribed. It is DNAase, an enzyme that destroys or "chews up" DNA. Dead white blood cells and bacteria in the airways make mucus thicker as they release their DNA. Pulmozyme, administered by nebulizer, destroys DNA in the airways and thins the mucus. It is a very expensive medication because it is manufactured by recombinant technology, a method that genetically engineers cells that can be grown in culture to produce this particular protein.

### INDICATIONS FOR USE OF MUCOLYTICS:

Mucolytics are indicated for pneumonia, bronchitis, emphysema and bronchiectasis when the disorder causes excessive thick, sticky pulmonary secretions.

### COMMON SIDE EFFECTS:

Many mucolytics can cause bronchospasm, which is when the muscles surrounding the bronchial tubes constrict and obstruct airflow. To minimize bronchospasms, it is recommended that a beta-2 fast-acting bronchodilator be used just before the mucolytic or along with the mucolytic, depending upon the decision of your healthcare provider.

Other potential side effects associated with mucolytics are nausea, runny nose, and coughing up blood. Although a few patients with thick sputum may benefit from mucolytics, the overall benefits seem to be very small. Therefore, the widespread use of these agents cannot be recommended for Alpha-1 COPD on the basis of the present evidence.

## EXPECTORANTS VERSUS COUGH SUPPRESSANTS

Expectorants and some cough suppressants can be purchased over-the-counter without a prescription; however, there are many different kinds of "cough syrups" available on the market, and deciding what type of product to buy can be quite confusing. Knowing the difference between these two types of medications can help you decide which form of medicine might be helpful for you. Expectorants work by thinning the mucus that blocks the air tubes leading to the lungs, and can improve your ability to cough up mucus or to "expectorate." This can be an important benefit for conditions such as pneumonia that tend to produce a lot of excess mucus. In these situations, coughing is helpful and should not be suppressed, because it helps to clear the airway of mucus build-up and makes breathing easier. When you can effectively cough, you will tend to cough less often, and this reduces the irritation to the lungs and throat. Because these products are swallowed and are not directly administered to the lungs, the scientific rationale for taking these products is somewhat controversial. Still, some individuals seem to gain benefit from expectorants, possibly from the soothing effect they produce more than the pharmacologic action associated with them.

One of the most common ingredients in cough medicine is guaifenesin. Guaifenesin is also available in pill form, such as Humibid. It is classified as an expectorant; however, some debate exists about how effectively guaifenesin does this. In addition, some cough medicines contain other ingredients that may cancel out guaifenesin's effects. Cough suppressants such as codeine, for example, work against guaifenesin because they discourage coughing-up the secretions that the expectorant loosens.

The easiest way to thin excess mucus in the lungs is to drink plenty of water. Dehydration increases the thickness and "stickiness" of mucus and works against effectively being able to cough up secretions.



**KEY LEARNING:** The easiest way to thin excess mucus in the lungs is to drink plenty of water.

Cough suppressants simply lessen the urge to cough. They should NOT be used if the cough is wet sounding and produces mucus. If your cough is hacking and does not produce mucus, however, a cough suppressant may be useful to minimize coughing, which in turn aids comfort and promotes your ability to sleep better. Some cough suppressants contain alcohol; therefore, it is especially important to follow the label instructions to avoid overuse and the possible side effects of excessive drowsiness and dehydration. Once again, drinking plenty of water is an effective way to reduce the irritation associated with a dry, hacking cough.

The most common over-the-counter cough suppressant is dextromethorphan. If more potent cough suppression is required, your physician may prescribe a cough syrup or pill containing codeine, a narcotic medication usually used for pain relief.

If you are considering purchasing an expectorant or cough suppressant, we recommend you consult with your healthcare provider to see which of these products might be right for you.

## ANTIBIOTICS

When antibiotics are used for individuals with Alpha-1 COPD, it is generally during an episode of acute exacerbation. Exacerbations are episodes of increased shortness of breath, usually accompanied by increased sputum production and increased purulence (yellow/green color of sputum).



**IT'S A FACT:** Antibiotic treatment is the mainstay of therapy for all acute exacerbations among Alphas, whether or not a bacterial organism has been identified as the causative agent.

Many people have been cautioned against overuse of antibiotics because it can lead to the development of bacteria resistant to common antibiotics. In addition, since exacerbations are not always directly caused by bacterial infections, the use of antibiotics in the usual treatment of COPD is somewhat controversial. Alphas, however, frequently develop bacterial infections from the damage caused to the lung tissue from the large influx of white blood cells that collect in the lungs as a response to viral infections or other types of irritants. Therefore, antibiotic treatment is the mainstay of therapy for all acute exacerbations among Alphas, whether or not a bacterial organism has been identified as the causative agent.

There are many antibiotics available to treat lung infections, and, in general, it is not clear if one antibiotic is necessarily any better than another. The types of oral antibiotics (pills) prescribed generally fall within one of the following categories: quinolones, macrolides, cephalosporins, sulfurs, and penicillins. To reduce the likelihood of bacterial resistance, in general, the same type of antibiotic should not be prescribed over and over again.

The organisms most commonly isolated from sputum in an exacerbation include: streptococcus pneumoniae (pneumococcus), hemophilus influenzae, and moraxella catarrhalis. In addition, recent evidence suggests that exacerbations of COPD are often caused by certain kinds of bacteria known as mycoplasma and chlamydia-type organisms, but these organisms cannot be cultured by routine methods.



**KEY LEARNING:** To reduce the likelihood of bacterial resistance, in general, the same type of antibiotic should not be prescribed over and over again.

Physicians often prescribe one of two classes of antibiotics (macrolides and quinolones) for people with chronic lung disease, because these types of antibiotics have been shown to be effective against infections caused by mycoplasma and chlamydia. Sometimes, your healthcare provider may ask you to produce a sputum specimen (that is, to cough up some phlegm) to perform a "culture and sensitivity" test. In this test, the organisms found in your sputum are allowed to grow in a laboratory dish. Different kinds of antibiotics are then applied to the "cultured" organism to see which ones appear to be the most effective at inhibiting the bacterial growth.



When one or more antibiotics are shown to be effective against the organism, it is said the organism is "sensitive" to these specific antibiotics. This testing process can be somewhat time consuming, and many times doctors will prescribe a "broad-spectrum" antibiotic (one that has been shown to be effective against a wide array of organisms) while they await the "culture and sensitivity" results.

All individuals need to be aware that allergies to antibiotics are somewhat common and typically are diagnosed if a rash or hives develop. Some allergic reactions to antibiotics can be life-threatening, and, therefore, you should always tell your healthcare provider about any medication allergies you may have. If you have an allergy to one member of a class of antibiotics, you also may be allergic to other members of the same class.

Other common side effects with antibiotics, in particular for macrolides and quinolones, include gastrointestinal symptoms such as nausea, vomiting, diarrhea, and stomach pain. Many of these symptoms can be alleviated if the medication is taken with food. Some Alphas have reported that they find it helpful to take plain yogurt and acidophilus when they take antibiotics.



**BURNING ISSUE:** Some allergic reactions to antibiotics can be life-threatening, and, therefore, you should always tell your healthcare provider about any medication allergies you may have. If you have an allergy to one member of a class of antibiotics, you also may be allergic to other members of the same class.

Other antibiotics often prescribed are listed below the table on the following page. Only antibiotics available in pill form have been included in these lists. During severe infections, it is sometimes necessary to administer antibiotics intravenously or, more rarely, intramuscularly. There are several classes of antibiotics not mentioned here that are only available for IV administration.

#### A PARTIAL LIST OF MACROLIDES AND QUINOLONES

TRADE NAME (EXAMPLES)	GENERIC NAME	FORM*
<b>Macrolides</b>		
Ery-Tab, E.E.S., Erythrocin	erythromycin	Oral or IV
Biaxin	clarithromycin	Oral
Zithromax	azithromycin	Oral or IV
<b>Quinolones</b>		
Cipro	ciprofloxacin	Oral or IV
Levaquin	levofloxacin	Oral or IV
Avelox	moxifloxacin	Oral or IV
Floxin	ofloxacin	Oral
Maxaquin	lomefloxacin	Oral or IV
Tequin	gatifloxacin	Oral
Zagam	sparfloxacin	Oral or IV

\*Oral = pills, capsules, liquids

\*IV = intravenous infusion

## OTHER ANTIBIOTICS COMMONLY USED INCLUDE:

- Penicillins: penicillin, amoxicillin, ampicillin, Augmentin.
- Cephalosporins: almost too numerous to mention, but prominently include Keflex, Duricef, Keflin, Velosef, Ceclor, Ceftin, Lorabid, Cefobid, Suprax. Rocephin is a frequently prescribed injectable used for exacerbations.
- Sulfur drugs: Bactrim, Septra, Sulfatrim, Cotrim.
- Tetracyclines: not usually recommended in this setting, but may be useful in some specific cases.

## INFLUENZA THERAPIES

The best therapy for infection by the influenza virus or "the flu" is prevention. This is accomplished primarily by obtaining an annual influenza vaccine or flu shot.



**KEY LEARNING:** It is recommended that FluMist® (the live-virus, nasal flu vaccine) not be used in Alphas with lung disease or in family members and other people who have close contact with Alphas with lung disease.

Unfortunately, although influenza vaccine reduces your risk of coming down with the flu, but it doesn't eliminate the risk entirely. In addition, there are times when, in spite of your best intentions, you forget your vaccination, or the strains of influenza covered by the vaccination are not quite correct.

If you do get the flu, there is now a form of therapy available that combats influenza virus. These therapies work in much the same way as an antibiotic kills bacteria. Currently there are two such products on the market, oseltamivir (Tamiflu) and zanamivir (Relenza). Relenza is available only in an inhaled powder form and is not recommended for individuals with lung disease.



**KEY LEARNING:** Relenza® is available only in an inhaled powder form and is not recommended for individuals with lung disease.

Therefore, this discussion of influenza therapies will be limited to Tamiflu, which is available as a capsule as well as a liquid for pediatric use. Each capsule contains 75 mg of drug. Tamiflu has been shown to be effective as preventive therapy following exposure to flu or as a treatment after symptoms of flu begin.

The effectiveness of this therapy is highly dependent on how soon after your flu exposure you start taking the drug the sooner the better! In any event, it is most effective if dosing is started within 48 hours after your exposure to the flu or within 48 hours of the start of your symptoms.



**KEY LEARNING:** In individuals with underlying lung disease, the prevention and treatment of influenza infection is such an important aspect of care patients should insist on receiving this specific therapy if their healthcare provider makes the diagnosis of flu within 48 hours of the onset of symptoms.

The dosing of Tamiflu is important.

- If you have the symptoms of a flu infection, Tamiflu should be taken twice a day for five days
- If you have been exposed to the flu and have not had an effective vaccination at least two weeks prior to the exposure, Tamiflu should be taken as a once-a-day dose and continued for at least a week

## ALTERNATIVES TO TRADITIONAL MEDICINES

Medication decisions require evaluation of the risks and benefits associated with each medicine, as well as how they may potentially interact with one another. You may hear claims about the potential health benefits of numerous herbal and vitamin supplements, or of homeopathic medications. These "complementary" or "alternative" therapies have not undergone the kind of rigorous clinical testing and review process required of prescription medications. This makes it difficult to objectively evaluate their health claims, and almost impossible to assess potential risks. There is a general sense among the patient community that because these medications are often offered in "health food stores" and over the Internet, it means there are not associated risks. This may not be the case.

Caution needs to be observed whenever "alternative" remedies are used. Since these supplements can be sold without FDA approval, they are not required to have standardized dosing units. For example, 200 mg of a specific supplement manufactured by one company may not be equivalent to 200 mg of the same supplement manufactured by another company. In fact, there can be considerable variations in the potency of the dosing units for different batches of supplements produced by the same manufacturer. In addition, alternative remedies may have undesirable interactions with some of the medications your physician prescribes.

More reliable information is beginning to emerge with regard to some complementary and alternative therapies. This information includes both better documentation of benefits and more complete understanding of the risks associated with taking these therapies. We encourage you to discuss all your medication usage, including prescribed and "alternative" formulations, with your healthcare provider. In this way, the combination of medications and supplements you intend to include in your treatment regimen can be evaluated in terms of what works best for you.



**KEY LEARNING:** Caution needs to be observed whenever "alternative remedies" are used. Since these supplements can be sold without FDA approval, they are not required to have standardized dosing units.

There is little disagreement that taking a multivitamin supplement may be beneficial to the general population, and especially for those who are stressed by disease. More controversial is the use of high-dose vitamin supplements. This is especially worrisome for the fat-soluble vitamins A and D, since overdose of these can be extremely dangerous.

An understanding of the role of oxidants in Alpha-1 lung disease has led some patients and physicians to consider the use of supplementary vitamins with anti-oxidant properties. These vitamins include vitamin C, vitamin E, and vitamin A. A typical regimen that would be expected to have minimal risk would include 500 mg of vitamin C and 400 units of vitamin E, taken morning and night. High doses of vitamin A can be toxic to the liver, and you should seek the advice of your healthcare provider before taking vitamin A to see if you can safely take this nutritional supplement. As with any medication, the regular use of any type of nutritional supplementation should be discussed with your healthcare provider.



**CROSS REFERENCE:** For more information on the use of supplementary vitamins with anti-oxidant properties see the Section about "Nutrition for All Alphas" in the Big Fat Reference Guide™ at [www.alphanet.org](http://www.alphanet.org).

MEDICAL INFORMATION WORKSHEET

CLIP & KEEP IN WALLET

Lined area for medical information.

LIST ANY SURGERIES (INCLUDING DATES)

Surgeries list area.



ADAPTED FROM THE ALPHA-1 DISEASE MANAGEMENT AND PREVENTION PROGRAM  
BIG FAT REFERENCE GUIDE™  
FOR ALPHAS, THEIR FAMILIES, AND THEIR DOCTORS

MEDICAL INFORMATION CARD



NAME

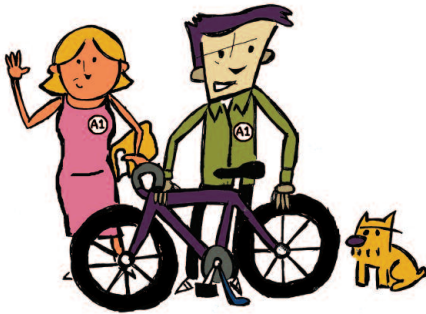
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MEDICAL INFORMATION WORKSHEET

CLIP & KEEP IN WALLET

NAME OF MEDICATION		
DOSE	FREQUENCY	
INHALERS AND NEBULIZED MEDICATIONS		
PILLS FOR LUNG PROBLEMS		
PROLASTIN OR OTHER ALPHA-1-SPECIFIC THERAPIES		
MEDICATIONS FOR OTHER CONDITIONS		
MEDICATIONS YOU HAVE AT HOME FOR AN EXACERBATION (E.G., ANTIBIOTICS, STEROIDS)		
OXYGEN THERAPY		
LITERS PER MIN.		

MY DIAGNOSES:
PRIMARY PHYSICIAN (NAME AND PHONE):
LUNG SPECIALIST (NAME AND PHONE):
EMERGENCY CONTACT PERSON:
NAME OF CLOSEST HOSPITAL:
DATE OF LAST FLU SHOT:
DATE OF LAST PNEUMONIA VACCINE:
DATE OF HEPATITIS B VACCINE:
DATE OF HEPATITIS A VACCINE:
ALLERGIES (ESPECIALLY TO MEDICATIONS):



This brochure is produced by AlphaNet as part of its Alpha-1 Disease Management and Prevention (ADMAP) program.

AlphaNet is a not-for-profit organization providing disease management services and support to individuals affected by Alpha-1 through a staff of medical professionals and specially trained AlphaNet Coordinators, available 24 hours a day, 7 days a week. To learn more about ADMAP or to find the AlphaNet Coordinator nearest you, visit our website ([www.alphanet.org](http://www.alphanet.org)).