Staying Healthy
Management of Environmental Risk Factors

Skinny Little Reference Guide™
INTRODUCTION

Alpha-1 usually does not, in and of itself, cause disease of the lungs or other organs. Rather, Alpha-1 makes individuals more susceptible to the risk factors that have the potential of causing organ damage in anyone. Thus, while certain environmental exposures might lead to relatively mild forms of lung or liver disease in individuals without Alpha-1, in those with Alpha-1 the same exposures can lead to more severe disease. It is important to appreciate that many individuals with Alpha-1 never develop any of these diseases. While some of this variability may be related to each individual’s total genetic makeup, the major influences on the development of disease appear to be exposure to risk factors that are entirely or partially controllable.

GOOD NEWS:
The major influences on the development of disease for Alphas appear to be exposure to risk factors that are entirely or partially controllable.

This brochure will help identify these risk factors and suggest ways that you can modify or reduce your exposure to them. Because this brochure deals with factors that you have direct control of, you have the opportunity to take steps to prevent disease or slow its progression.

For individuals who are “carriers” of a single abnormal Alpha-1 gene, reduction of risk factors deserves special attention. Although the risk of developing diseases may be lower for carriers when compared to Alphas with two abnormal genes, there is a growing appreciation that carriers are also at increased risk for disease development. Therefore, controlling your exposure to these same risk factors is just as important for you.
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THE FACTS ABOUT SMOKING

Nearly 500,000 people in the United States die each year from smoking cigarettes. Smoking is a well-known cause of cancer, stroke, heart disease, COPD and peripheral vascular disease, and exerts harmful effects on many other systems in the body as well. Recent long-term studies indicate that approximately one-half of all regular cigarette smokers will eventually die from their addiction.

Since having Alpha-1 already puts you at an increased risk for developing lung disease, the damage inflicted by smoking will almost guarantee it. Therefore, your first priority for managing the risk factors associated with developing lung disease in Alpha-1 should be the elimination of exposure to all forms of tobacco smoke.

THE EFFECTS OF SMOKING IN ALPHA-1 ANTITRYPSIN DEFICIENCY

Smoking affects every part of the body. Cigarette smokers inhale over 400 toxins and 43 known carcinogens every time they puff. As smoke enters the respiratory tree and lungs, it causes irritation and triggers inflammation. This inflammation causes the body’s defenses to send white blood cells to the area. While performing their normal function, the white blood cells release a powerful enzyme, known as neutrophil elastase. Neutrophil elastase is destructive to unprotected lung tissue. Alpha-1 antitrypsin (AAT) is the protein in our bodies that provides protection to the lungs by neutralizing this powerful enzyme.

For the smoker, inhaling cigarette smoke is known to accelerate the destructive processes at work in the lungs and puts the development of lung disease on the “fast-track.” For Alphas who smoke, that fast-track becomes an “express train.” But it is not only the risk to the smoker that is of concern. Secondhand smoke can also cause considerable damage to the lungs of Alphas who do not smoke themselves.

In addition, children of smokers appear to have a much higher incidence of asthma than children of non-smokers. If your child is either an Alpha or an Alpha-1 carrier, the risk of developing lung disease in later life will be greatly increased by exposure to secondhand smoke. The risks associated with cigarette smoke, especially in the presence of Alpha-1, are clear and profound.

Although cigarette smoking is recognized as a controllable risk factor, it is clear that, in many cases, it is not an easily avoidable one. Some Alphas live with smokers who are unwilling to quit. Although some Alphas never take another puff of a cigarette after they are diagnosed, this is certainly not always the case. It is important to understand that dealing with a cigarette addiction, as with any addiction, may be very difficult. Successful change takes knowledge, help from others, and a long-term commitment to health.

Since this brochure is all about knowledge, let’s start with some facts.
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SMOKING IS THE GREATEST RISK

First among the controllable risk factors associated with development of lung disease in Alpha-1 is exposure to cigarette smoke. Whether from personal use or secondhand exposure, cigarette smoke has been undeniably shown to exert the greatest risk.

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IT’S A FACT: A single cigarette destroys all the alpha-1 antitrypsin in the lungs.
WHY QUIT?
Despite knowing the harmful effects of smoking, smoking cessation can be a challenging and difficult undertaking because it involves overcoming physical dependency and nicotine addiction, as well as psychological factors. Nevertheless, the benefits of quitting are numerous, significant, and undeniable. Some of these benefits include:

- Decreased airway inflammation
- Prevention of the inactivation of alpha-1 antitrypsin
- Decreased morbidity — you'll live longer
- Decreased risk of heart disease, lung disease, and cancer
- Decreased risk of other health effects
- Improved lung function — less shortness of breath
- Slowed rate of decline of lung function
- Increased energy
- Smoker’s cough goes away
- Digestion normalizes
- Liver function normalizes
- Healthier environment for those who live with you, especially children
- Freedom from the mess, smell, and burn holes in clothing and furniture
- More money to spend on other things

KEY LEARNING: Although smoking cessation may be one of the most difficult lifestyle changes you make, it is the most important step you can take to improve and preserve your quality of life. Stop now.

DEVELOPING A SMOKING CESSATION PLAN
There are numerous resources available for individuals who want to quit smoking.

- Organizations such as the American Lung Association and the American Cancer Society offer smoking cessation programs.
- Your local hospital and your health care provider can provide you with treatment options and a list of smoking cessation programs in your area.
- There are also countless resources on the Internet, for those who have access to this valuable tool.

GOOD NEWS: Research has shown that the harmful effects of smoking on the liver are temporary and most problems can be reversed when the individual stops smoking.

We know the chemicals created from burning tobacco destroy alpha-1 antitrypsin. In fact, a single cigarette destroys all the alpha-1 antitrypsin in the lungs. Because each cigarette smoked results in a repeated episode of irritation and inflammation, this process may therefore account for a significant portion of the lung disease seen even in non-Alphas. Among individuals with normal AAT levels, this damaging process generally occurs gradually. The symptoms of lung disease in these individuals, if they occur, tend to develop during their 50s or 60s.

For Alphas, where protection against neutrophil elastase is already compromised by reduced AAT levels, the exposure to cigarette smoke increases the risk that lung damage will occur and will result in significant symptoms of lung disease. As opposed to “normal” individuals, this damaging process is accelerated in Alphas and their symptoms may develop as early as in their 30s.

In view of the relationship between smoking and AAT levels, individuals preparing to receive augmentation therapy for Alpha-1 should be smoke-free and successful in maintaining smoking cessation before therapy is initiated. This makes sense if you remember that “augmentation” therapy is simply giving you a “boost” of AAT. If you smoke, the AAT boost will be destroyed just as your own AAT is destroyed by cigarette smoke.

While almost everyone is aware of the links between smoking and heart and lung disease, few people think about the effects of smoking on other parts of the body. When the various toxins and products of cigarette smoke enter the lungs, they subsequently enter the bloodstream, where they circulate to the remainder of the body. One of the liver’s primary functions is the processing of drugs, alcohol, chemicals, and other toxins to remove them from the body. Evidence suggests that smoking alters the ability of the liver to handle and “detoxify” such substances. Some research also suggests smoking can aggravate the course of the liver disease caused by excessive alcohol intake. Despite this damage, additional research has shown that the harmful effects of smoking on the liver are temporary and that most problems can be reversed when the individual stops smoking.
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GOOD NEWS: Research has shown that the harmful effects of smoking on the liver are temporary and most problems can be reversed when the individual stops smoking.
The first step is to have the desire and motivation to quit. For some, the diagnosis of Alpha-1 is the primary motivation. Once the decision is made, discuss developing a smoking cessation plan with your health care provider and your family and friends. Enlist their help and support. If other members of your family are smokers, encourage them to join you in quitting. The importance of maintaining a smoke-free environment within the home should be stressed and promoted. A few steps towards being smoke-free are:

**ALTERNATIVE SMOKING CESSATION METHODS**

Acupuncture is being more widely used but has yielded variable results, and it may be too soon to know whether or not this approach will ultimately prove to be of benefit. Behavioral techniques, such as hypnosis and counseling have been of benefit for some, as has participation in smoking cessation programs and support groups. In fact, combining more than one form of treatment may increase your chances of success.

**IT’S A FACT:** For many, quitting will not be easy. If it were easy, there would be far fewer smokers in the world!

**IF AT FIRST YOU DON’T SUCCEED ...**

Most smokers will have tried to stop on several occasions with varying degrees of success before they finally “kick the habit” for good. Smokers need to be constantly encouraged by their physician and family members. The physician’s active participation in the process is invaluable. Ask your friends or family for their support. They may want to stay in touch with you to offer ongoing support. If you should suffer a lapse and return to smoking, then it’s back to the starting point: set a date, make a plan, and put the plan back into action.

Of the various smoking cessation strategies, none have been shown to be consistently more effective than another, but one may succeed where another has failed. So if you are not successful the first time, don’t give up! Seek help and support. If you resume smoking after having stopped for a period of time, evaluate where your difficulty arose and consider utilizing nicotine replacement therapy, behavioral intervention, or individual or group counseling to help improve your chances of success.

**CROSS REFERENCE:** A list of resources available to help you with your smoking cessation efforts can be found in the Big Fat Reference Guide, at www.alphanet.org.

Many Alpha-1 Aware physicians report that when they discuss the cold, hard facts of smoking and its damaging effects on the body with their newly diagnosed Alphas, as many as 90 percent are able to quit without further intervention. This is particularly true for individuals who have already developed respiratory symptoms.
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I DON’T SMOKE — WHAT ARE OTHER RISK FACTORS?
Risk factors affecting individuals with Alpha-1 may be found in the home, workplace, or outdoors. As you review the information presented here, consider the factors that may pose the greatest risks for you. If you suspect that some of the elements we identify could be responsible for producing your symptoms and ill health, then implementing a simple plan to reduce or eliminate your exposure may offer relief and promote an improved sense of well-being.

Each individual has a unique situation and set of circumstances. Not all of the recommendations presented here may be useful to you. Keep in mind that no environment, outside of living in a bubble, is 100 percent risk-free. Despite this fact, there is no need to live in constant fear. In general, it is the long-term and repeated exposures to irritating and toxic substances that have the greatest health consequences.

RISK FACTORS RELATED TO LIVER DISEASE
Risk factors for the liver disease of Alpha-1 are not as well identified as those for lung disease. The liver is the largest organ in the body and is vital to keeping the body functioning normally. It removes toxins from the blood, including chemicals, germs, and bacteria. It manufactures the protein alpha-1 antitrypsin and other proteins necessary for blood clotting. The liver also plays a role in digestion and utilization of nutrients, because it produces bile that helps absorb fats and vitamins.

It is believed that substances that can be toxic to the liver in “non-Alphas” may have increasing toxicity in individuals with Alpha-1. These liver toxins can contribute to, or compound, the liver damage that Alphas may develop, and can lead to cirrhosis, liver cancer, and/or liver failure.

Chief among the substances known to cause liver damage is alcohol. Excess alcohol consumption is by far the most common cause of toxic chemical damage to the liver in our society as a whole. The American Liver Foundation defines excessive alcohol consumption as any amount greater than two drinks per day. Other agents that are inhaled, swallowed, or absorbed through the skin can also cause damage to the liver. Among these are solvents, toxic alkaloids, pollutants, dietary supplements, and prescription and over-the-counter drugs.

Many different drugs have been implicated as a potential cause of liver damage. The widely used pain medication acetaminophen (Tylenol®) is one such drug. Acetaminophen is generally safe when taken as prescribed. However, when taken in excessive doses, either all at once or over a period of time, acetaminophen can cause severe damage to the liver. In individuals who regularly consume alcohol in excessive amounts, acetaminophen can be toxic even at lower doses.

RISK FACTORS RELATED TO LUNG DISEASE
Numerous substances that can affect the lungs, including smoke, asbestos, radon gas, and bacterial and viral agents, have been extensively studied and their harmful effects are well documented. A number of environmental factors, including industrial smoke, toxic fumes, solvents, dust, and gases, have been shown to be associated with the development of lung disease or with aggravating existing respiratory conditions. Because our lungs are continually in direct contact with elements in the air, it is especially important for individuals predisposed to breathing difficulties due to Alpha-1 to recognize potential risks and learn how to safeguard themselves. By first recognizing the risks and limiting exposures, harmful effects can be minimized or avoided altogether.

HOME ENVIRONMENT
Aside from tobacco smoke, other potentially harmful substances encountered in the home environment include:

- Fumes
- Aerosolized products
- Powders
- Dust and dirt
- Bacteria
- Mold and mildew
- Household cleaners
- Bug sprays
- Fireplace smoke
- Asbestos
- Radon gas

There is no conclusive evidence showing a direct relationship between the development of specific lung diseases and exposure to any of the common substances found in the home. However, individuals who are sensitive to substances such as cleaning products, soaps, deodorants, and cosmetics may experience respiratory difficulties with exposure. Individuals with Alpha-1 or those with known or suspected allergies may experience difficulty breathing when exposed to pet dander, mold, mildew, pollen, or other airborne particles. Persistent exposure by sensitive individuals to any of these elements may lead to long-term respiratory problems.

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RISK FACTORS IN THE HOME, AT WORK, AND OUTDOORS

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Various sources of potentially harmful agents in the home are identified below, along with simple strategies to eliminate or reduce the risk of exposure.

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<th>HARMFUL AGENTS IN THE HOME</th>
<th>HELPFUL TIPS</th>
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| **FUMES** generated in the kitchen, bathroom, workroom or garage, from unvented kerosene heaters, gas heaters, and construction or building materials, specifically: formaldehyde from particle board, fiberboard, and hardwood, plywood, wall paneling, textiles, carpets, and carpet padding. | • Institute the use of a mask whenever the potential for breathing harmful or irritating fumes exists.  
• Open windows and increase ventilation when you are in areas where fumes may accumulate or install exhaust fans. Remember to inspect fans and clean them routinely.  
• When working on your car in a garage, keep the garage door open and prevent exhaust fumes from entering your home.  
• Keep lids secure on solvent containers. Dispose of saturated cloths or rags in sealed containers. Always use a mask and gloves when handling these substances to avoid or reduce inhaling the fumes and protect your skin from contact.  
• Have heaters inspected and serviced regularly. Check pilot lights on gas heaters and stoves to make sure the gas flame is burning blue, not yellow or orange.  
• Use “exterior grade” pressed wood products because they emit lower amounts of phenol resins as opposed to urea resins.  
• Assure that carpets and carpet padding have little or no formaldehyde content. |
| **AEROSOLIZED** products may include deodorants, hair sprays, perfumes, air fresheners, and spray disinfectants. | • Use roll-on deodorants, pump sprays, solid air fresheners, and non-toxic alternatives. |
| **POWDER** is readily airborne and can be inhaled, causing irritation and potential respiratory problems. | • Avoid use or powder sparingly to avoid breathing airborne particles. If you must use powder, consider using a mask. |

**HOUSEHOLD CLEANING/INSECTICIDE PRODUCTS**, ammonia-based products, petroleum-based products, oven cleaners, bleach, spray furniture polish, bug sprays, paint sprays, and mildew remover sprays. Use cautiously as these products pose a threat to your liver, as well as your lungs. Remember, the liver detoxifies what we breathe.

- Minimize your use of toxic chemicals. Seek less toxic or more natural alternative products as much as possible. For example, baking soda or a solution of vinegar and water serve quite well as household cleaning products.
- Use commercially available pre-moistened dusting/polishing cloths or a cloth slightly dampened with water.
- Use extra caution when using chemical sprays – make sure the room is well ventilated and wear a mask.
- Be aware of the nature of chemicals that come in contact with your skin, because these can be absorbed through the skin and affect your liver. The insecticides used for trees and shrubs not only kill bugs, but can be absorbed through the skin and damage the liver. That is why they are called toxic chemicals. Use gloves, a mask, hat, and protective clothing every time you handle these substances. Protect your skin from exposure as much as possible.
- Never mix chemical solutions, because deadly chemical reactions can occur. For example, never mix ammonia and bleach!
- If irritating chemicals must be used, arrange for someone else to do the cleaning and leave the home until the fumes have dissipated.

**DUST AND DIRT** can accumulate quickly and cause breathing difficulty.

- Avoid all dusty situations, including shaking rugs, vacuuming, sweeping, and dusting. Have someone else do the cleaning if possible. If you must be the one who does the dusting, wear a mask.
- When cleaning areas where dust and dirt routinely accumulate, use a damp rag or mop to reduce airborne particles. Remember, many household appliances collect dust and other irritants and should be cleaned regularly as well. These include dryer filters, refrigerator coils, drip pans, heat exchangers, furnaces, heating ducts, and vents. Also, central air conditioning and heating filters should be replaced regularly. You may wish to consider the use of a HEPA (High Efficiency Particulate Air) filter in your home.
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### HARMFUL AGENTS IN THE HOME

| FUMES generated in the kitchen, bathroom, workroom or garage, from unvented kerosene heaters, gas heaters, and construction or building materials, specifically: formaldehyde from particle board, fiberboard, and hardwood, plywood, wall paneling, textiles, carpets, and carpet padding. | • Institute the use of a mask whenever the potential for breathing harmful or irritating fumes exists.  
• Open windows and increase ventilation when you are in areas where fumes may accumulate or install exhaust fans. Remember to inspect fans and clean them routinely.  
• When working on your car in a garage, keep the garage door open and prevent exhaust fumes from entering your home.  
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### HELPFUL TIPS

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- • When cleaning areas where dust and dirt routinely accumulate, use a damp rag or mop to reduce airborne particles. Remember, many household appliances collect dust and other irritants and should be cleaned regularly as well. These include dryer filters, refrigerator coils, drip pans, heat exchangers, furnaces, heating ducts, and vents. Also, central air conditioning and heating filters should be replaced regularly. You may wish to consider the use of a HEPA (High Efficiency Particulate Air) filter in your home.
A NOTE ABOUT GEOGRAPHIC LOCATIONS

Many individuals with Alpha-1 are concerned about how a specific geographic location will affect their overall health, and some question if they should move to a different location. There are areas around the country that are known to have poor air quality and these exert a harmful effect on health. If you are concerned about the air quality of a particular location, the American Lung Association has a dedicated Internet site (www.lungusa.org, click on “air quality”) that is a helpful resource for reviewing air quality for each state and county.

Individuals with Alpha-1 lung disease are likely to experience more difficulty breathing in locations where the altitude is significantly higher than sea level. This is because the concentration of oxygen is less at higher altitudes than it is at lower altitudes. This reduced oxygen concentration forces the lungs to work harder and places a strain on a respiratory system that is already compromised.

The consideration of whether you can live “at altitude” will depend on your functional capabilities at that altitude and whether or not breathing symptoms are present and tolerable. Currently, there is no evidence to suggest Alphas without lung disease can prevent the development of disease by changing their place of residence from a higher altitude to a lower one.


DUST AND DIRT can accumulate quickly and cause breathing difficulty.

HEPA filters are the highest efficiency air filters available for the filtration of small particles. Defined by the Institute of Environmental Science, a certified HEPA filter must capture a minimum of 99.97 percent of contaminants at 0.3 microns in size.

FIREPLACE/WOOD BURNING/STOVE SMOKE pose risks for individuals with respiratory difficulties. Although economic factors and the availability of resources may influence the decision to use one type of heating method over another, consideration should be given to the overall impact of this method of heating on your respiratory health. Choosing an alternate method may be indicated.

• Provide for cleaning of the chimney on an annual basis, or more frequently if needed.
• Before lighting the fireplace, make sure the damper is open. For a wood stove, be sure it is in proper working condition, the seams are tight and sealed, and the gasket is not overly worn and creating gaps.
• Burn only firewood, not paper, charcoal, or other items that can cause toxic fumes.

BACTERIA, MOLD AND MILDEW can accumulate in areas in the home that are moist, damp, and dark, especially bathrooms, kitchens, basements, garages, and humidifying and dehumidifying units. The soil of house plants is also known to support the growth of mold and mildew.

• Increase ventilation in bathrooms and kitchens with fans vented to the outdoors. Wash tile and grouted surfaces frequently. Regrouting may be needed occasionally.
• Wash and replace sponges used in the kitchen and bath frequently, especially during the cold and flu season. Launder them in the washing machine with bleach or soak them in a bleach solution. Replace soiled hand towels with fresh ones routinely.
• Seal leaks and waterproof basements. Wipe up any leaks or standing water as soon as possible.
• Clean and dry water-damaged carpets, or remove and replace them altogether.
• Consider growing house plants in an enclosed terrarium.

ASBESTOS AND RADON GAS health hazards are well documented. Asbestos can be found in old deteriorating insulation, fireproofing material, or acoustical materials. Radon gas occurs naturally in the earth, in uranium and rocks beneath homes, old granite foundations, and in well water.

• Have qualified and trained contractors handle the control of asbestos exposure, removal, and/or clean up.
• Have your home tested for radon and seek professional assistance before planning abatement measures.
• Seal cracks and openings in basement floors and walls. Provide for adequate ventilation in the crawl space. Treat radon-contaminated well water through aeration or filtering through activated charcoal.

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Humidity may also be a factor that affects breathing for some Alphas. Some individuals report that they are able to breathe more easily in a humid location, while others prefer a dry climate. Without further research, one geographic location cannot be recommended over another for maintaining good health because each individual’s response and tolerance is different.

Before making any dramatic changes in your geographic location for the sake of your health, we suggest you visit the area you are considering for relocation and arrange a prolonged visit. You may want to visit during the various seasons to assess how seasonal changes may affect you. Consideration should be given to the prevalence of allergens and other irritants that may be present in a specific location as well. Before instituting any changes, you may wish to review them with your health care provider to evaluate if the change is a good choice for you.

**WORK ENVIRONMENT**

A variety of substances in the workplace cause health hazards. Exposure to them can pose serious risks for healthy individuals, as well as present increased risks for those with Alpha-1. It is important for individuals diagnosed with Alpha-1 to be aware of these risks and make appropriate occupational choices when possible. This is especially important for younger Alphas who are just starting out in their careers and can make more informed career and vocational choices. Choosing “Alpha-friendly” work environments and avoiding occupations that repeatedly expose you to toxic substances can reduce your risk of developing lung or liver disease.

For those who receive their diagnosis while working in an occupation that places them at constant risk, consideration should be given to job reassignment, vocational counseling, retraining and/or new employment.

Toxic substances in the workplace present themselves in varying physical forms such as smoke, gas, dust, liquids, vapors, or mists. The amount of exposure will affect the extent of damage sustained. Many substances pose dual risks for individuals with Alpha-1 because both the lungs and liver are vulnerable. While some hazards pose an immediate threat to health and life, others produce damage and ill effects after repeated and longstanding exposure. An example of a disease that develops after long-term exposure is silicosis, a serious lung disease seen in workers who are repeatedly exposed to silica dust. Another example is the development of hepatitis after repeated exposure to carbon tetrachloride among individuals who work in a confined space without protection of a respiratory mask.

In the presence of inadequate ventilation, breathing hazards can become readily visible as clouds of dust or fumes or detected as strong odors. These substances can also irritate the eyes, nose, and throat. You should keep in mind that some harmful substances have no odor, such as carbon monoxide. Also remember your sense of smell can become accustomed to odors after repeated exposures, and you may no longer be aware of it — even though the danger may still exist.

The following is a listing of some common toxic substances found in the workplace, along with examples of their specific effects:

- **AMMONIA**: A gas that is an irritant to eyes, nose, and throat, and causes upper airway constriction.
- **ASBESTOS**: A dust that causes lung fibrosis or scarring of the tiny air sacs in the lungs and surrounding tissue. Also causes lung cancer.
- **PHOSGENE**: A toxic gas that causes pulmonary edema, chemical pneumonitis, and corrosive burns to air passages. Also causes death of liver cells.
- **CARBON MONOXIDE**: A colorless, odorless gas that interferes with the ability of the blood to carry oxygen to the vital organs of the body.
- **CARBON TETRACHLORIDE**: A colorless liquid that is a known carcinogen and toxic to the central nervous system, liver, and kidneys.

When work environment risks are identified, action should be taken to eliminate or avoid exposure to the source. It is your employer’s responsibility to keep you informed of the general and more specific hazards associated with your employment and provide you with a safe work environment.

**BURNING ISSUE**: If your job places you in a situation where you are repeatedly exposed to toxic substances, your employer is required to provide you with appropriate safety equipment including respirator masks and protective clothing.

As the employee, it is your responsibility to be knowledgeable about the substances you will come in contact with in the workplace. Know where the Material Safety Data Sheets (MSDS) are kept and what the recommendations are for avoiding, reducing, and eliminating exposure, as well as immediate care in the event of exposure. It is also your responsibility as an employee to report an unsafe and unhealthful work environment to the Occupational Safety and Health Administration (OSHA).
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The use of masks or respirators, outside of those required and regulated in occupational environments, may be an option for individuals who want to reduce their exposure to potentially harmful substances found in the air.

**A Respirator** is a device that protects you from inhaling dangerous substances, such as chemicals and infectious particles. The term “mask” refers to any device that minimizes your exposure to airborne infectious diseases and other potential irritants such as dust, dander, mold, fumes, chlorine, printer’s ink, ozone, particulate matter, sulfur dioxide, formaldehyde, gasoline, natural gas, and smoke. For some individuals, minimizing exposure to breathing cold, “icy” air can also be achieved by the use of a mask.

**CROSS REFERENCE:** Additional information regarding numerous other types of specialized masks can be found in the “Additional Resources” section in the Big Fat Reference Guide, at www.alphanet.org.
Respirator masks are designed to reduce the risk of exposure associated with inhaling smoke, dust, fumes, mist, or particles. Respirator masks are very specific for the substance being used or the particle size of the elements that might be inhaled. Employees must have an appropriate mask for their individual use and have it fitted and tested to specification. There can be no gaps or leaks that allow contaminants to enter and risk unwanted exposure. A mask with a leak, or one that is worn-out or damaged, should not be used; it should be refitted and retested.

For individuals with existing breathing difficulties, a respirator mask may not be acceptable because the mask itself can impose breathing restrictions. In this case, the inability to safely wear protective equipment may require job reassignment away from the breathing hazard. You should work closely with the occupational health nurse, your supervisor, and physician to facilitate this process. The possibility of seeking a new work environment, alternate employment, or even disability may need to be considered.

OUTDOOR ENVIRONMENT
Poor outdoor air quality and air pollution affects everyone, but individuals with pulmonary problems are at greater risk. Individuals with liver problems may also be at risk. Contaminants are present all around us from varying sources: exhaust from vehicles, discharges from industrial plants, air pollution, smog, chemicals, fertilizers, pesticides, bacteria, mold spores, and fungi. All of these substances contribute to poor air quality. As previously mentioned, certain geographic locations are known to have poorer air quality than others. The American Lung Association can provide further information regarding your particular geographic area.

Despite taking precautions, you may still experience breathing difficulties when exposed to pollutants and other irritants. Always consult your physician before you adjust your medications or the level of your supplemental oxygen. Your health care provider is the individual who is best able to assess your condition and offer you the appropriate recommendations for treatment and risk avoidance.

AIR POLLUTION
In communities where air pollution can be routinely problematic, reports and warnings are broadcast to the public during news reports and weather updates. Depending on the level of air quality, warnings and instructions are given as to whether it is advisable to be out of doors and what level of activity is appropriate. Adhering to the recommendations during periods of poor air quality and instituting some simple measures can reduce your risk of exposure.

HEALTHFUL TIPS
• Avoid poor air and noxious fumes when traveling by car. Steer clear of traveling on crowded highways at the height of rush hour. Whenever possible, seek alternate routes away from congested highways or plan travel when traffic is lighter. Keep the air conditioner on and roll up the windows.
• Stay indoors when air quality reports deem it necessary. Keep windows closed and turn on air conditioners and air filters.

THE USE OF MASKS
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Some individuals with existing respiratory problems may find breathing through a mask difficult. However, there are many models to choose from, and you may be able to find one that best suits your individual needs and situation.

Here are some simple tips to keep in mind when choosing a mask and for assuring a proper fit:
- Determine the substances or conditions from which you desire protection and what mask is most effective for those specifications.
- Is the mask reusable, washable or disposable? Follow the manufacturer’s handling and cleaning procedures.
- Do not allow the mask to become wet or soiled. If this occurs, replace it.
- The mask should fit securely over your nose and mouth creating a seal. Check for unwanted gaps.
- Men with beards may have to shave or trim facial hair to achieve a good seal.
- The use of oxygen via nasal cannula may prevent achieving a tight seal.
- Do not allow the mask to obstruct your vision. Adjust the nose clip, if applicable.
- Adjust the straps for a snug, comfortable fit.

The use of masks can be an option in promoting your personal safety by reducing the potential for exposure to irritating and harmful substances in the air. If you have an understanding of the need for protection and the types of masks available, you can choose appropriately for maximum protection and benefit.

MINIMIZING THE RISK OF INFECTION

Of all the environmental hazards individuals face, the most fearsome enemies remain the microorganisms — bacteria and viruses. Airborne diseases causing acute respiratory infections are responsible for the deaths of four million people worldwide per year, and are the leading cause of death among children under the age of five. Viral infections affecting the liver are reported to cause between 6,000 and 10,000 deaths annually in the United States alone.
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GOOD HAND-WASHING TECHNIQUE

1. WET YOUR HANDS with WARM running water.
2. ADD SOAP AND THEN RUB your hands together, away from the running water, making lather. Continue for at least 20 seconds using a good amount of friction, rubbing the front and back of your hands, between your fingers, and under your nails. Concentrate on your knuckles and don’t forget your wrists.
3. RINSE YOUR HANDS WELL under warm running water. Let the water run back into the sink.
4. DRY YOUR HANDS THOROUGHLY on a clean towel, preferably a paper towel that can be easily disposed. Use the paper towel to shut off the faucets.

Be sure to wash well with soap and water as soon you are able. Have waterless soap or wipes available when access to soap and running water is limited. Products containing alcohol with a concentration of at least 60 percent may be useful, but washing with soap and water is still preferable.

GOOD ADVICE: To limit the risk of infection while away from home, take along a supply of antibacterial wipes, sprays or other products to clean surfaces with which you will necessarily come in contact. Items such as seatbelts and tray tables on airplanes, and phones and light switches in hotels can be sources of bacteria. Remember however, that your best protection against infection is to wash your hands. If clean water is unavailable using the antibacterial products is the best way to guard against infection.

Note: If hands or surfaces are visibly soiled with organic material (sputum, feces) handiwipes and waterless soaps are not effective because they do not remove the material, but instead, merely smear it around. Soap and water should be used in these instances.
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THE ONE-TWO PUNCH

Individuals with Alpha-1 are more susceptible to respiratory infections and the consequences of infectious agents affecting the liver. To reduce the risk of experiencing serious complications that may be associated with these potentially fatal infections, vaccinations against influenza, pneumonia, and hepatitis are recommended for all individuals with Alpha-1.

FLU VACCINE

The influenza virus causes influenza or “the flu.” Infected droplets that are inhaled from the air spread the flu from person to person. Unlike other viral respiratory infections, such as the common cold, the flu can cause severe illness and life-threatening complications for many people. Approximately 36,000 people per year in the United States die from influenza, and 114,000 per year are admitted to the hospital as a result of the flu.

GOOD ADVICE: A yearly flu vaccination is recommended for individuals who are at increased risk for serious complications from the flu including individuals who are age 50 and older, individuals with chronic pulmonary or heart disease, persons having a compromised immune system, residents of nursing homes, and health care workers.

Symptoms of flu include fever, headache, fatigue, cough, sore throat, runny or stuffy nose, and muscle aches. Children may have nausea, vomiting, and diarrhea, but these symptoms are uncommon in adults. Complications of the flu can occur and include bacterial pneumonia, dehydration, and worsening of chronic medical conditions, such as congestive heart failure, asthma, or diabetes. Individuals with chronic medical conditions or who are 65 years of age or older are at the highest risk for serious complications from the flu.

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### MINIMIZING THE RISK OF INFECTION

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According to the National Advisory Committee on Immunization (NACI) and the ACIP, a one-time re-vaccination with Pneumovax®-23 is currently recommended for the adults:

- Who are at the highest risk for developing a fatal pneumococcal infection
- Who did not experience a severe reaction to the initial vaccination
- Who were previously vaccinated with Pneumovax more than five years ago

Re-vaccination is not required for individuals who were vaccinated less than five years ago.

HEPATITIS VACCINES

Hepatitis is inflammation of the liver that can lead to its swelling, tenderness, and scarring. The most common types of viral hepatitis are hepatitis A, hepatitis B, and hepatitis C. Viruses that are transmitted cause the disease from various sources including contaminated food or water, oral-fecal route (hepatitis A), or through contact with infected blood or body secretions (hepatitis B and C).

Hepatitis B and C may become chronic conditions that can lead to serious, permanent liver damage and death. It may be advisable to immunize Alpha-1 individuals with vaccines against hepatitis A and hepatitis B, because there is some evidence that having Alpha-1 can make these viral infections of the liver worse.
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**HEPATITIS VACCINES**

Hepatitis is inflammation of the liver that can lead to its swelling, tenderness, and scarring. The most common types of viral hepatitis are hepatitis A, hepatitis B, and hepatitis C. Viruses that are transmitted cause the disease from various sources including contaminated food or water, oral-fecal route (hepatitis A), or through contact with infected blood or body secretions (hepatitis B and C).

Hepatitis B and C may become chronic conditions that can lead to serious, permanent liver damage and death. It may be advisable to immunize Alpha-1 individuals with vaccines against hepatitis A and hepatitis B, because there is some evidence that having Alpha-1 can make these viral infections of the liver worse.
GOOD NEWS: With proper oral care, gum disease can be controlled or even reversed.

The human mouth always contains some bacteria, but the presence of dental infections of any type increases the number of bacteria present and can also lead to the growth of bacteria that are particularly hard to treat if they cause infection in the lungs or airways. With proper oral care, gum disease can be controlled or even reversed.

WHAT CAN I DO?
Visiting your dental hygienist on a regular basis is one of the most important steps you can take to maintain or improve your oral health. Your dental hygienist will review your medical history, clean and polish your teeth, and refer any areas of concern to your dentist or physician.

Developing a good daily cleaning regime, along with routine office visits with a dental hygienist, will control or reverse gum disease. Less than five minutes, twice a day, is all it takes to maintain or improve oral hygiene:

BRUSHING: Place your brush at a 45-degree angle to the junction between tooth and gum, applying gentle pressure as you move the brush away from the gums. Don’t forget to brush your tongue (with or without toothpaste), where bacteria build up. You should brush about three minutes each time you brush.

EASY AS PIE: Less than five minutes, twice a day, is all it takes to maintain or improve oral hygiene.

FLOSSING: Wrap 18 inches of floss around your middle fingers until you have a two-inch length between them. With the thumb and forefinger of each hand, guide the floss gently and carefully between each tooth in a “C” shape and gently guide it up and under the gum line.

Antibacterial mouthwash: In addition to the above, some advocate the regular use of antibacterial mouthwash, particularly during cold and flu season.

DENTURES: Even if you wear dentures, it is still important to clean your mouth and get regular check-ups to prevent oral health problems.

GOOD ADVICE: It may be advisable to immunize Alpha-1 individuals with vaccines against hepatitis A and hepatitis B, because there is some evidence that having Alpha-1 can make these viral infections of the liver worse.

ORAL HYGIENE AND OVERALL HEALTH
We all know prevention is one of the keys to maintaining good health. Taking good care of our dental health is another important step in maintaining overall health.

The reasons for maintaining good oral hygiene are much more than cosmetic. While at one time it was believed the worst outcome of gum disease was tooth loss, studies have shown oral health affects the entire body. Bacteria that are present in the mouth, particularly from decaying teeth or infected gums, can easily enter the bloodstream and migrate throughout the body causing damage to the heart, lungs, and other organs.

Bacterial respiratory infections are usually thought to be acquired when fine droplets in the air are inhaled. These droplets contain germs that can breed and multiply within the lungs. While this is clearly one way lung infections arise, recent research suggests the bacteria found in the mouth and throat can be drawn into the lower respiratory tract and cause infections in the lung and bronchial tubes.

Individuals with poorly functioning immune systems, the very young and very old, and Alphas who have some degree of lung disease suffer from reduced protection against such infections, making them more susceptible to respiratory infections. Good oral hygiene is, therefore, a very important component of a disease management and prevention program, especially for these individuals.

WHAT IS GUM DISEASE?
According to some estimates, as many as 75 percent of adults over the age of 30 may suffer from some degree of gum disease. Gum disease begins with the formation of hard and soft deposits on the surface of the teeth. Over time, a build-up of bacteria, called plaque, collects at the gum line, eventually hardening on the teeth into calcium deposits called calculus or tartar. With poor oral care, these bacteria can cause inflammation of the gums, or gingivitis, penetrate the gum line, and finally spread into the underlying bone.
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SUMMARY

While having a genetic condition brings with it certain uncontrollable risks and predispositions, for Alpha-1, many of the important risk factors are, in fact, controllable and manageable. The conscious choices you make in your environment can safeguard your health. This section has described the known risk factors for disease in Alpha-1 and suggested ways to reduce these risks. The changes needed to protect your health often involve cooperative efforts between you, your family, your employer, and your health care providers. This cooperation is important to maximize your continued health and quality of life.
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