Cruising on. Next Stop... Adulthood

Successful Strategies for Adolescents and Young Adults with Cystic Fibrosis

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Dear Teen and Young Adult with CF

Question:

How do you write a booklet that you know will give teens and young adults with CF important information on life and moving forward into adulthood compelling and fun to read?

Answer:

Tough job! We want this booklet to be useful in helping you "live well" with CF! We want to provide you, the adolescent & young adult with CF some helpful hints on how to do that. We have tried to make the information relevant to your life so that you will want to read it. It is our belief that this booklet will give you some very helpful information. But, is it fun and compelling to read? Well, you be the judge of that!

We start out with material that tells you what happens to every teenager and how CF can affect that. We then talk about how to become skilled at taking great care of yourself and CF. Next are tips for handling some of the tough stuff CF throws at you, and finally, how to look ahead and plan your future. Check out the resource section at the back of the booklet. There are some helpful tools for you to copy and use. We also include a list of helpful websites you may find useful.

So, hold on to your hat and start reading. Keep in mind that it might not be the most fun read ever, but it may be the most important read you've read so far about CF.

Sincerely,

Mary Jo McCracken

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Next Stop… Adulthood
Successful Strategies for Adolescents with Cystic Fibrosis

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I. Tasks of Adolescence in Relation to Cystic Fibrosis

Adolescence is a time of great change. It is all about moving from being a child to growing into adulthood. That is a pretty remarkable transition! With that transition, almost everything changes. Your body changes dramatically in size shape and function. Your brain also changes significantly in its ability to problem solve, see things more abstractly and look beyond the present and into the future.

Socially, things also change as you move away from your family and become more independent, more focused on your friends and begin dating and developing intimate relationships. When adolescence begins and ends varies from person to person but usually takes place anywhere between 10 to 21 years of age.

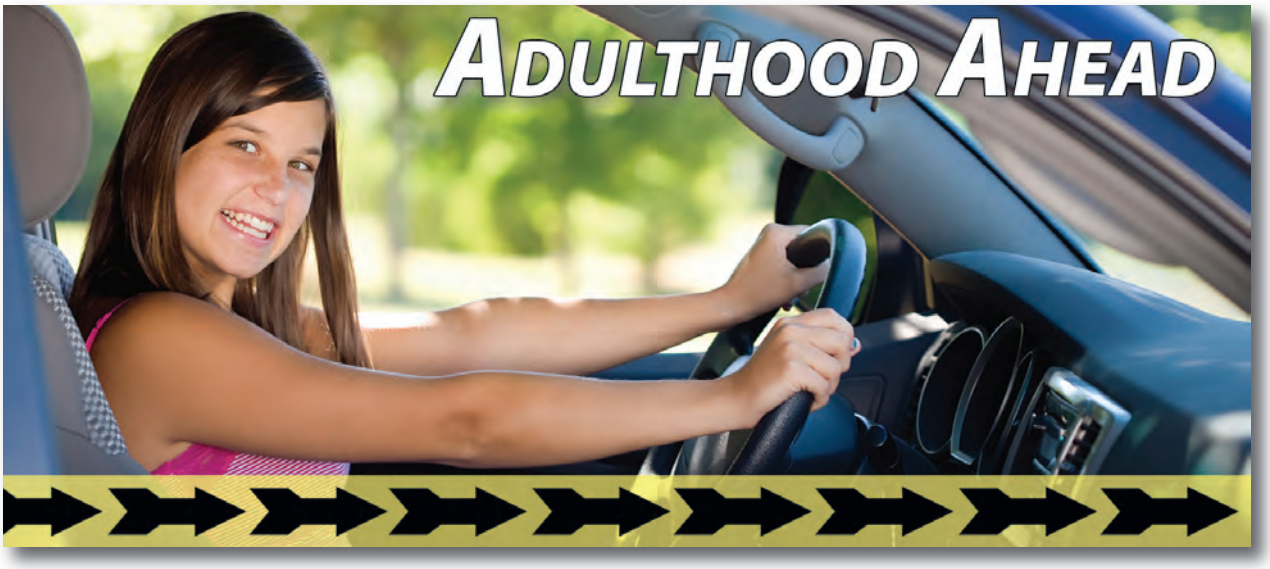
During this time there are several "tasks of adolescence" or certain developmental changes that all adolescents experience as they work their way toward adulthood. How and when these changes take place can be different for teens like you who have cystic fibrosis (CF).

How CF can affect the changes that take place during adolescence, the developmental tasks and what you should be aware of is the focus of this first chapter. As you go through this section, keep in mind that you have some control over what and when many things happen during your teenage years. You are the driving force; don't forget that!

The purpose of this booklet is to help you make this journey somewhat easier - to give you the knowledge and skills to take hold of your life and rise to the challenges you might encounter. Putting yourself in the driver's seat, (not CF) is the core theme of this booklet. Knowledge is power, and the hope is that by educating you on some of the ins and outs of adolescence and CF, you become more powerful to take charge of your life. That is the mission!

A. Physical Growth

The bodily changes that happen during adolescence can be compared to those that happen to a newborn baby during its first year of life. The body's size and shape are dramatically different from beginning to end. Both boys and girls have significant growth spurts, girls before boys, and usually reach adult height around 18 years of age. This remarkable growth is going to require a whole lot of fuel (food) for this to happen. It



is true, we are what we eat. During the teenage years nutrition is critical. The growth that takes place during adolescence requires enormous energy (which is calories) and using all this energy is a lot of work. This is why teenagers sleep so much!

For teens with CF who are pancreatic insufficient and require digestive enzymes with all meals and snacks, getting enough calories in each day to support this growth can be a major challenge. Lung disease with infections and inflammation affects appetite and burns up many calories making it even more challenging. For these reasons, many males and females with CF who historically have had difficulty growing and gaining weight can lag behind others their age in physical growth and development. They can be thinner, shorter, less developed and generally younger looking. Males can be about one and a half years behind in experiencing the physical changes of puberty. For females, the onset of menstruation is generally two years later than for girls without CF.

B. Forming Identity

Developing a sense of who you are and how you relate to others is what psychologists call your "personal identity" and it is an important task of adolescence. Every teen goes through times when they question, "Who am I?" and "Why am I here?" Inner turmoil and anxiety are common. For teens with CF these feelings can be more intense as you see the differences that can come with CF. Once again keep in mind that you have control over how you see yourself and who you become. If you see yourself as weak and sickly, that is what you will become. However, if you see yourself as someone who is strong, creative and can rise above some limitations, that is what will happen.

Developing friendships and spending time with friends plays a big role in how teens develop and mature. If you are not physically able to participate in highly active sports, take up less strenuous activities where you can use your creativity and other skills to develop friendships and spend time away from home. Remember there are many ways to interact with peers and friends. Student council, debate team, drama or chess club, the school paper or yearbook, and music are just a few of the activities one can participate in. It is important to get involved and make new friendships. That is part of the fun of being this age.

Risk taking and rebellion are typical ways adolescents test their limits and forge their own identity. For teens with CF, this behavior can often be directed at things that can significantly hurt them and even have life-threatening consequences.

Refusing medications (like digestive enzymes when eating) or airway clearance techniques (ACT)* are common. Some even question whether they really have CF. Smoking and abusing chemicals (alcohol or street drugs) will have a negative impact on your health. If one could step outside their body and look objectively at their behavior, they would see quite quickly just how stupid those behaviors are. "Why would one want to hurt oneself?" you might ask. Hopefully, you can be mature and think through this behavior. When under intense peer pressure to smoke, take drugs or participate in other actions you know are not a good idea, use your CF as a convenient, really good reason not to. Use lines like, "My CF doctor would kill me if he/she found out!" or "My CF doctor told me I could get really sick if I did that!" This is probably one of the few times you might be glad you have CF.

** ACT (airway clearance techniques).* These are treatments that loosen the thick, sticky mucus in the lung so it can be cleared by coughing and huffing. This reduces lung infections and improves lung function. They include chest physical therapy (cpt), the Vest, positive expiratory pressure therapy (PEP), bronchial drainage (BD), acapella® (oscillating PEP), the Flutter, active cycle of breathing (ACBT), autogenic drainage (AD). For some patients with CF, it also includes aerosol inhalations (nebulizer treatments).

C. Interpersonal Relationships and Sexuality

Sexuality, intimate relationships and reproductive health are significant issues for all adolescents and young adults. Those with CF are no exception. As mentioned earlier, physical development can happen at a slower rate in CF but it does come, and catch-up growth occurs. Sexual growth and development are major changes during adolescence and are fueled by excellent nutrition. When breast development and menstruation begin will depend on your genetics (what age your mother was when she began menstruation) and nutritional status.

CF does not affect the female reproductive organs and females with CF are able to have children. It is a common misconception that they can not. Some women with CF may have difficulty conceiving a child because of thick mucus that lines the cervix, but others do not. It is important that lung function and nutritional status are at acceptable levels before a woman with CF becomes pregnant to help facilitate a healthy pregnancy. A frequent question is whether ACT (airways clearance techniques), especially the vest, will hurt the unborn fetus. The answer is "no."

Women with CF should talk to their healthcare provider (HCP*) about birth control and which method is right for them before becoming sexually active. They need to remember they can become pregnant and that this should be a well-planned, happy occasion, not an unexpected, unwanted surprise. Women with CF are frequently on antibiotics to treat lung infections and need to be aware that many antibiotics can make birth control pills ineffective and, therefore, additional protection should be used when sexually active. It is advisable that sexually active men and women practice safe sex (protecting yourself from undesired pregnancies and sexually transmitted diseases).

Frequent antibiotics can kill the "good bacteria" in a woman's vagina, which can result in a vaginal yeast infection. A yeast infection usually causes a thick discharge which has a strong odor and is itchy. These may be difficult to treat when antibiotics are used often or for long periods of time. They can be eliminated, but for some it may take perseverance and patience.

A female problem for 25 percent of women with CF is urinary incontinence (leaking urine) with coughing or laughing. This is also common in women after having a baby. It is highly treatable and women need to discuss these issues with their CF HCP* so they can be treated. Don't let embarrassment keep you from getting these feminine problems resolved.

Sexual maturity for males with CF also depends on their nutritional status. Most males with CF are infertile, but this, in no way, affects the ability to have sex, which is totally normal. The infertility is the result of a blockage in the vas deferens, which is where sperm need to travel to meet the semen. It is similar to having a vasectomy. It is now known that many men with CF make sperm but it is trapped in the testes which is in the scrotum. Today the technology exists to retrieve trapped sperm and artificially inseminate their partner so men with CF can father their own children. Again, sexually active men and women need to practice safe sex.

Prior to having children, all partners of adults with CF should be checked to see if they are carriers of the CF gene. Cystic fibrosis is caused by a recessive gene, which means that in order for a child to be

* **Healthcare provider (HCP)** refers to the person you see for your health care needs. This could be a physician or nurse practitioner. (CF HCP) refers to the person you see for your CF healthcare at the CF Center.

born with CF, it must receive a CF gene from both the mother and the father at conception. All individuals with CF will pass a CF gene to their unborn child making them carriers of the CF gene (just like your parents). Whether that child will have CF will depend upon whether the partner has a CF gene to pass on. If they do not carry a CF gene, then their children will not have CF. If the partner does carry the CF gene, there is a 50 percent chance with each pregnancy that you have with this partner that the child will have CF. Consult a genetic counselor when considering having children.

D. Independence

The struggle for independence and control is another well known task of adolescence. It is the source of numerous power struggles that take place between adolescents and their parents and other authority figures. Your need for independence may differ from your parents' views as you get older. Parents of teens with CF are at risk for sending mixed messages about independence, in part because of their own concerns about the future. It is these concerns that lead some parents to become overprotective. This can lead to more rebellion and frustration for the teen. The overprotective parent can send a strong message to the teen which is "you can't do it; you won't succeed" when the opposite is true; you can do it, you just have to go about it the right way.

Seldom do teenagers, regardless of whether they have CF, move from complete dependence on parents (or other adults) as children, to complete independence as adults. It is more realistic to work toward a goal of "inter-dependence" as you work your way to adulthood. You will need to sit down with your parents and decide how they can help you with your CF as you get older. Taking on more responsibility for your CF cares* will help get nagging parents off your back. The next chapter on self-care will be especially helpful in this area.

Some teens with CF find it difficult to tell friends that they have CF. Unlike childhood friends who have grown up knowing you have CF, there is concern that learning about your CF can affect new friendships. How can it affect these friendships? Friends will need to understand and accommodate how much time and effort must be spent taking care of yourself. They will also need to understand some of the physical complications such as decreasing lung function, lower energy level and the impact on quality of life. Also, they will need to know that CF is generally life limiting. Not everyone needs to know all there is to know all at once, but the longer one goes not mentioning CF, the harder it can become. The best approach for many is not to make it a big deal and simply be factual: "By the way, I have a health condition called CF. I have to spend time each day doing lung treatments and take some medicine each time I eat so I can use my food better." Not much more needs to be said and you can decide if you want to answer questions or share more, but now you don't have to worry about it anymore. For most people you tell, it won't make any difference at all. For the few it might, it is good to know that early in the friendship and it may be better not to have them as a friend after all.

Similarly, as you develop close, romantic relationships with a boy-or girlfriend, you will need to discuss how and when they can provide support and assistance with your CF. That may mean coming to your CF clinic

visits or reminding you to do your ACT. Whatever it is, make sure they are things that you would find helpful, not nagging or detrimental.

A major source of rebellion for adolescents with CF is their CF cares. Adherence to the CF care program is a challenge for individuals with CF and their families at all ages, but adolescence proves especially difficult. It is unlikely that you will wake up one morning and say, "I am now ready to manage my CF cares completely." The transition to self-care is a gradual process that has several steps necessary for success. Chapter II is all about self-care and how you and your family should go about it so you avoid some of the battles and pitfalls commonly experienced by other CF families.

E. Cognitive Development

The brain is also developing during the adolescent years. This is called "cognitive development." You are now able to go beyond thinking in concrete and absolute terms and begin to think "outside the box." This means you can now start to think in abstract, theoretical, or speculative ways. This enables you to solve complex problems and think about your future and how to plan for it. The life-threatening complications of CF become a reality and thoughts about death and dying occur. Worrying about the future and what adult life will be like is common. "Will I be able to work and support myself, live alone, get married, have children?" Most teens with CF welcome the opportunity to talk about these feeling and fears. They just need to be asked. Some are angry that despite hearing about scientists working toward finding a cure for CF through much of their childhood, such a cure has yet to be found. Strong feelings from a variety of things can lead to high-risk behaviors such as cigarette smoking or abusing alcohol and/or drugs as a way of saying they "know they will die young anyway." Intervention by CF and chemical abuse experts is essential during these times.

F. Planning for the Future

A key task of adolescence is looking to the future, setting goals and making plans for achieving those goals. The future holds great promise for further advances in the fight against CF. Individuals with CF need to sit down with their CF HCP and CF social worker to set goals for their future, including realistic educational and vocational goals. High-school guidance counselors can also be helpful.

Planning for the future also involves moving your CF care from the pediatric CF clinic you have been attending to the adult CF center. Different CF centers have different plans for moving older teens and young adults to their adult CF centers. Although it is painful to leave the CF team who has cared for you for years, (sometimes since you were a baby) this is a move that indicates that you are now an adult and it's time you were treated like one! The adult CF team members are specialists in CF as well as in adult problems and issues and will be able to follow you for decades. That is nice! The last chapter of this booklet deals specifically with the future in more detail.

* **CF cares** refers to all the things one with CF does to take care of their CF. It includes medications, airway clearance techniques (ACT), digestive enzymes, high-calorie diet, cleaning home equipment and any other prescribed cares.

II. Self-Care for the Adolescent with Cystic Fibrosis

A. What Is Self-Care?

Self-care is perhaps the most important subject in this booklet. What's so special about self-care? Well, self-care is your road map, or ticket, to your dreams! And you should have BIG dreams for your life. None of this "I can't" stuff because of CF. You can, and will, be able to fulfill your dreams if you learn how to take care of yourself and keep yourself healthy. That's the wonderful thing about self-care: It opens the door to so many possibilities.

So, what does self-care really mean? Self-care simply means taking care of yourself – GOOD care of yourself. For individuals who live with a chronic illness, self-care means putting some thought into how your body is working each and every day. If you do this, and do it well, you have a greater chance of feeling good, being healthy and doing the things you want to do today, tomorrow, next week, next month and in the future. Therefore, self-care is your ticket to everything: health, independence, success, getting nagging parents off your back, going away for the weekend, going away to college, getting your own apartment, traveling and everything you might dream of doing. (Add your own dreams to this list.) A world of possibilities opens up for you when you become self-caring. Create your own dreams list and keep it in an obvious place.

B. What Do You Need for Successful "Self-Care"?

1. Knowledge

The first step for successful self-care is **knowledge**. Knowledge is actually the foundation for self-care. You need a rock-solid understanding of CF and how it affects the body for you to make the best health care decisions, which are a big part of self-care. Even though you have lived with CF your entire life, there may still be some aspects of the disease you don't understand. Now is the time to learn all about CF. The good news is that there are many resources available to teach you about CF. Your knowledge needs to be comprehensive because you are the leader of your CF team. This means YOU are going to be responsible for making health-related decisions. To do a good job, you had better know what you are doing!

There is an excellent tool called the *CF Self-Care Questionnaire* (see the Resource Section, Table I) which was developed to measure one's knowledge of CF in self-care. Fill it out and see what you need to learn about CF in order to become the expert you need to be. Once you have taken the *CF Self-Care Questionnaire*, compare your answers to the Answer Key (Table II) which follows the Questionnaire (Table I) in the Resource Section. This should show you which areas in CF you need to understand better. Again, keep in mind that the decisions you make about your healthcare will be based on how well you understand CF and its treatments.

Researchers and scientists are always learning new things about CF and how it affects the body. Keeping up with new information through newsletters, the Internet, CF conferences, etc., will be very important for advancing your CF knowledge and your ability to make good decisions for yourself. Your CF center can help you with this.

There are also many web sites that provide excellent information on CF, especially the Cystic Fibrosis Foundation web site at: **www.cff.org**. Table VI in the Resource Section at the back of this booklet lists additional websites you may find informative.

2. Performance Capabilities

The second step needed for successful self-care is **performance capabilities**. This means doing your CF cares the way you are supposed to and correcting any bad habits you may have developed. For example, do you adjust (increase or decrease) the number of digestive enzymes you take with different meals and snacks? Do you correctly prepare and take your aerosol treatment and perform airway clearance techniques as prescribed by your CF HCP each day? Or, do you do your Vest treatment as you were instructed or do you play computer games the whole time and forget to change frequencies and cough or huff? Do you increase the number of ACT's you do each day when you have a chest cold and/or a significant change in your sputum (thick and green)? Are you cleaning your aerosol equipment correctly every day? All of these questions are part of self-care and your performance capabilities.

It is a good idea to make an appointment with your CF team members at the clinic and have them assess your performance capabilities. Then, if needed, they can re-instruct you in areas that may need more attention. It is important to remember that if you are going to do your CF cares, take the time to do a good job that will be helpful, not to waste your time and certainly not to hurt yourself. How could being sloppy with your CF cares hurt you? There are several ways. Here are two examples:

First, if the nebulizer cup used for your aerosol treatments is not properly cleaned, bacteria will grow and you could inhale these bacteria directly into your lungs the next time you use it. This could lead to a serious lung infection. The second example involves not properly adjusting digestive enzymes when eating different foods. For example, you should increase the number of enzymes you take when eating high-fat foods. High-fat foods are higher in calories and therefore, are foods you should be eating. But they are also more difficult to digest so you will need to take more digestive enzymes when eating high-fat foods. Not taking or not adjusting your enzymes can result in several different complications including increased malabsorption and loose stools, weight loss, or over time constipation, DIOS* (distal intestinal obstruction syndrome), or the dreaded rectal prolapse*! This is a very strong argument for taking good care of yourself. It is best to remember to not only take but also adjust your enzymes with each meal; however, forgetting a few times should not cause DIOS or rectal prolapse*!

3. Support People

The third step for making self-care work for you is having **support people** available. Identifying individuals, both teens and adults, you can look to for strength and comfort in good times as well as challenging times, is important. For most teens, this naturally is their family: parents, brothers, sisters, aunts, uncles, cousins, grandparents. For others, it might be a best friend or a best friend's mother, a teacher, a coach or school nurse. It can be any person you can count on to boost you up, listen to you, help you problem-solve and generally be there for you when you need them. Support people change over time as we move and grow. There are usually more support people available to you than you think. Individuals are often very willing to play a role in your life. All you need to do is ask. Line up your support people in order of priority, starting with your most important and working down to the ones you might use for emergencies only. You may have school support people and then summer camp support people. The idea is to have support people where you might need them. Whatever works for you is what counts.

List your support people here:

_____

_____

_____

_____

_____

_____

_____

_____

** DIOS* is a partial bowel obstruction typically caused by thick mucus, dehydration and undigested food. If not treated, it can become a complete bowel obstruction, which is a medical emergency. DIOS in the past was called MECONIUM ILEUS EQUIVALENT.

** Rectal Prolapse* this is when the rectum protrudes or falls outside the anus. In CF, it is usually from maldigestion and malabsorption where large amounts of fatty, bulky stools overtime become too much for the rectal tissue and it prolapses. This can be a sign of CF in undiagnosed children. One is usually able to gently push the rectal tissue back into the anus. Rectal Prolapse can also occur if individuals with CF do not take, or do not take enough, of their digestive enzyme supplements.

4. CF Self-Care Teams

The fourth step for successful self-care is setting up your **CF self-care teams**. There will be two parts to this step. The first part includes your CF self-care teams. There will be three different teams: the **professional**, **personal**, and **PRN CF self-care teams**. The second part has to do with your role in all of this. Since it is your body we are talking about, your role will be the Team Leader or "Head Honcho." Let's get into your role as team leader first, since this is critical. Remember, **you** are the driving force; it is your body we are talking about. Then, once we are finished with your role, we will set-up your three CF self-care teams.

You as Team Leader

You will have four different jobs as Team Leader. The **first** and perhaps most important is assessment and decision-making when it comes to your health. As you develop more independence and knowledge in your self-care, this will become one of your primary jobs. It involves thinking about how your body is feeling each day in a systematic way (your **"body check"**), deciding if this is a change from how you usually feel (which is called your **"baseline"**) and if it is, do you need to do anything about it? Your body usually tries to let you know if things are not going as well as they should be. It gives you clues that something is wrong. You need to learn to listen to your body, to look for clues that something is not right. When you do listen, it gives you the opportunity to make some changes in your treatment plan that could prevent you from getting sick or prevent damage from happening to certain body parts. This is HUGE in CF where a lot of damage can be occurring without you really realizing what is happening. CF is a sneaky disease! It often takes a lot of that thick, sticky mucus full of bacteria plugging up your airways and damaging the tissue before you start to feel sick. So individuals with CF need to be smart and catch changes that are happening before real, irreversible damage occurs.

This is what the body check is all about. Learning to listen to your body so you can "nip problems in the bud" (my grandma would always say that!). It means taking care of things before real problems happen.

The body check doesn't have to take a long time. What you should do is develop a computer program in your head just for this job. You should start at the top of your head and work your way down checking each body part for how it is feeling and whether this is a change from your "baseline." Using a numerical rating system with your body check will make things clearer and easier to put it all together rather than "a little bit better" or "a little bit worse." Use 1 to 10 with 1 being the best you could ever feel, 5 being average and 10 being the worst ever.

For some of you, it may be helpful to write down (log) your body check. This will help keep things clear until you become an expert and are able to do this in your head. Table III in the Resource Section is a sample log sheet which will help you log your daily body checks. It is ideal to do your body check every day. If that seems impossible to do, then do it once a week. Don't throw the whole idea out just because you can't do the ideal. Doing it some of the time is better than not doing it at all.

Another way to use the body check and log sheet is for making your assessment of how you are doing and what has changed right before your CF clinic appointment. You can actually take the log sheet with you to share with your CF HCP. This can be a wonderful way to prepare for your clinic visit and get the most out of it. Your CF HCP will be delighted. The body check log sheet will give your CF Team a clear picture of how you are doing and where there might be problems.

There is a blank log sheet in the Resource Section (Table III) for you to photocopy and use when doing your regular body checks.

Throughout this next section there will be **"sidebars"** which are little bits of wisdom, CF information or advice that is pertinent to what is being discussed.

Sidebar

A good portion of your daily body check is probably best done in the morning during your ACT or right after it. It is a good idea to do ACT first thing in the morning, before breakfast. Why? When you sleep, your lungs sleep too, which allows a lot of thick, sticky mucus, and maybe lots of bacteria too, to pool and accumulate. Doing ACT with coughing and huffing will get that mucus/sputum out. Getting this out will make you feel better and have a better day (cough less in school, eat better, study better and have more energy). You can probably get a good portion of your body check done while you are doing this morning ACT.

The CF Body Check Log

Okay, so let's get started!

Use the rating scale below to answer the following questions:

At each bullet you will be asked to rate the status of the body part, as well as, answer different questions. *For each of these – circle the correct number or response.*

1	2	3	4	5	6	7	8	9	10
excellent	very good		good/average		fair		poor	sick	worst ever

First, how do you feel overall?

Circle your correct #: 1 2 3 4 5 6 7 8 9 10

How does this compare to yesterday? Is it better/ worse/the same? If it is worse, you will need to figure out why. You may discover why when doing the rest of your body check. Let's continue...

Check your appearance.

Circle your response:

Is there anything unusual or different about your face? (swelling, sunken eyes, rash, bruising)

How is your color? (pink, pale, dusty, blue-tinged lips)

Is this a change? (yes/no)

Do you feel feverish? (yes/no)
If yes, take your temperature. If it is above or at 101 degrees, call your CF clinic.

How did you sleep?

Use the rating scale to score your night's sleep:

Circle your correct #: 1 2 3 4 5 6 7 8 9 10

How does this compare to the night before? Is it better/worse/the same? Are you able to lie flat or does coughing, post-nasal drip or shortness of breath prevent you from doing so? Is this a change (yes/no)? If yes, do you need to do something about it? If so, what should you do?

Suggestions: call the CF clinic, do ACT with good huffing/ coughing before bed or raise the head of the bed when you sleep.

Upper Respiratory System

- **NOSE**

Do you have nasal problems? (nasal polyps, frequent congestion? (yes/no) If no, continue on to sinus question. If yes, answer the next few questions. Can you breathe through your nose? (yes/no) What is the status of your nose?

Circle your correct #: 1 2 3 4 5 6 7 8 9 10

Is this a change from yesterday? Do you have nasal secretions? (yes/no) If yes, what color are they (clear/white, yellow, green, blood-streaked)? Is this a change? (yes/no) Estimate the amount of nasal secretions you have had for the past 24 hours (____ tsp(s), ____ tbsp(s), ____ cup(s)? Is this more or less than yesterday (more/less/no change)? Do you need to do anything about the changes in your nose?

Suggestions: call the CF clinic/CF HCP, use nasal spray or allergy medications as directed, call ENT Clinic or Specialist.

- **SINUSES**

Are you having sinus problems? (yes/no) If yes, do they feel full, congested, any facial pain? (yes/no) What is the status of your sinuses?

Circle your correct #: 1 2 3 4 5 6 7 8 9 10

Is this a change from yesterday (yes/no)? If yes, what do you need to do about it?

Suggestions: call the CF Clinic/CF HCP, or ENT Clinic or Specialist use nasal spray, use other medications such as MOTRIN® for pain or inflammation, do nasal irrigations/wash if recommended by your CF HCP or ENT Specialist.

Do you have a headache? (yes/no) If yes, what is your status?

Circle your correct #: 1 2 3 4 5 6 7 8 9 10

Is this a change from yesterday? (yes/no) Do you need to do anything about the pain in your head?

Suggestions: take Motrin®, Tylenol®, call the CF Clinic/CF HCP. If you are on oxygen, you may need to have your blood gasses checked.

Sidebar

Headaches, especially upon waking in the morning, can be caused by carbon dioxide (CO_2) retention. This happens when an individual with CF has very serious chronic obstructive lung disease and the area to exchange gases (breathe in oxygen [O_2] and breathe out CO_2) is limited. During sleep, when secretions have accumulated and further decreased the lungs' ability to take in O_2 and breath out CO_2, the carbon dioxide builds up and causes a headache. Doing good ACT with coughing and huffing will remove these retained secretions and thereby "blow off" the CO_2 and improve the headache.

- **MOUTH**

Are your lips pink or bluish/dusty? (yes/no) Is this a change? (yes/no) Is the mouth free of sores, bad teeth or toothache? (yes/no) Is your throat red, swollen or sore? (yes/no) What is the status of your mouth?

Circle your correct #: 1 2 3 4 5 6 7 8 9 10

Is this a change from yesterday? (yes/no) Do you notice any abnormally foul breath or taste in your mouth? (yes/no) Do you need to do anything about these changes, if you have any?

Suggestions: call the CF clinic/CF HCP or dentist if you think you may have and infection and you may need antibiotics, or perform any prescribed mouth cares.

Sidebar

Any time you notice a foul taste in your mouth or foul odor to your breath that persists, even though you have brushed your teeth well and have used mouthwash, take notice! If this continues more than a day or two, call your CF clinic, especially if you have any other changes like increased cough or sputum production, change in sputum color or a low-grade fever. A foul breath/taste can often be a sign of an anaerobic infection in your lung (this is caused by bacteria that do not need O_2 to live, and give off a foul odor or taste as a clue to their presence). It could also be a sign of an infection in a problem tooth.

Lower Respiratory System

- **LUNGS**

The lungs are obviously the most important part to check for individuals with CF. There are several issues regarding the lungs that you will need to check.

Respiratory Rate: While at rest, start by counting the number of breaths you take for a full minute (don't count for 15 seconds and then multiply by four). **Remember, you need to be at rest for a while** when you count your respiratory rate since physical activity will increase it. You need to establish your "baseline respiratory rate." Generally, it is 12 to 16 breaths per minute for adolescents. When the body feels the need for more O_2, it will increase the rate of breathing. Physical activity, like running up stairs, will increase your rate of breathing as will congestion or more mucus/sputum in your lungs. Is your respiratory rate significantly different (by more than four breaths) from your baseline (yes/no)? If your respiratory rate has increased it may be a clue that something is amiss in your lungs. So be on the alert and monitor things closely.

Sidebar

Lung infections are a fact of life for most individuals with CF. Certain bacteria love the CF lung and problems with bacterial infections can start early in life. Treating these infections aggressively is key to controlling CF lung disease. When certain bacteria start to grow in the CF airway, a vicious cycle gets set in motion.

The lung recognizes bacteria as hazardous and begins to make more mucus in an attempt to clean the bacteria out of the lung. However, the lung doesn't recognize that there is a problem with the mucus in CF. The mucus is too thick and sticky and therefore is difficult for the cilia to move it upward. So the mucus begins to clog or obstruct the small airways in the CF lung. The bacteria are delighted with this because this gives them an opportunity to grow even more. Now the bacteria begin to damage the tissue of the airways. At first, the airways become swollen. This narrows the airway diameter making it even more difficult to get mucus up and out.

Then the cilia begin to get destroyed, thereby compromising the major mechanism for moving mucus up and out of the lungs. If left untreated, the bacteria will continue to damage and destroy lung tissue and can result in **Bronchiectasis**. This is where the airway is so damaged it is overly dilated, with no cilia or elastic fibers. This leaves mucus to stagnate and just sit there where it becomes a "glob of gunk or pus." The only way to get this "gunk" out is with ACT.

This is why it is so important to treat these infections and halt the destruction they can do to your lungs. Whether the

treatment is oral antibiotics, intervenous (IV) antibiotics or aerosolized antibiotics, it is critical to use these drugs as prescribed by your CF HCP so they can work.

• COUGH

Has your cough changed? (yes/no) If yes, how? How often do you cough (never, rarely, some days, most days, everyday, only with ACT or exercise)?

Frequency of cough? __None, __1-2 times every twelve hours, __1-2 times every 6 hours, __1-2 times every 2 hours, __1-2 times each hour, __many times every hour.

Is your cough productive (No sputum, clears throat/swallows, sometimes productive, always productive)?

What makes you cough (ACT, laughing, exercise, crying, colds, tickling, aerosols, cigarette smoke)?

What is the status of your cough? 1 (none), 2 (clears throat), 3-4 (mild), 5-6 (moderate), 7-8 (a lot), 9 (severe), 10 (worst ever)?

Circle your correct #: 1 2 3 4 5 6 7 8 9 10

Is this a change? (yes/no) If yes, what should you do about it?

Suggestions: call your CF clinic/CF HCP, increase the # of ACTs each day, watch closely for other changes, or increase huffing/coughing.

Sidebar

Do you suppress your cough? Many CF individuals of all ages suppress their cough until they learn how destructive it can be. Then the majority will change their behavior. If you suppress your cough you need to stop. Coughing is your body's way, or defense mechanism, to get rid of stuff it knows is not good. It is like a sneeze. If you hold back a cough, you are working against your body's natural ability to keep itself well. That is not very smart. Work with your body to actively cough and huff. Get that mucus and bacteria up and out of your lungs so they are clear and can breathe easier.

• SPUTUM

Sputum is a name for the mucus in your lungs. A change in the characteristics of your sputum can be very important. You should assess your sputum each day for the amount you cough up, the color and the consistency. How are you going to do this? It is very important to cough your sputum into a tissue and then look at it. Don't get grossed out! Your body is trying to tell you something. Listen to it. That is what you are doing when you look at your sputum. Any increase in the amount of sputum you are coughing up tells you that your lungs are being irritated by something and are attempting to get rid of this irritant through their self-cleaning mechanism. This mechanism, the muco-ciliatory escalator, involves making mucus to trap the irritant and other undesirable crud and sweeping it upward by cilia toward the throat, where it can be coughed up and out.

Amount: The amount of sputum is an indicator of how much irritation is occurring. An increase in the amount of sputum means things are getting worse and a decrease usually means things are getting better. In CF, the irritant is most likely bacteria that have caused an infection. Lung infections are common in CF and it is important to be aggressive in treating them to lessen the possibility of permanent lung damage. It is common to have a sputum or throat culture done at clinic to check for lung bacteria. Become knowledgeable about your culture results and signs and symptoms of infection. The irritants can also be a virus or an allergen. Both of these can cause the lung to make more mucus.

Sidebar

A time when less mucus is **not** a sign of improvement is with **bronchospasm**. Here the mucus is trapped and prevented from being coughed or huffed up and out of the lung by a spasm in the airways or bronchi. Bronchospasm occurs when some trigger (allergen, virus, infection, etc.) causes the muscles lining your airways or bronchi to spasm or clamp down. When this happens the diameter of your airways/bronchi gets significantly smaller and narrower making it more difficult for air to pass through. A **wheeze** is the sound made by air trying to pass through airways/bronchi in a spasm. If the bronchospasm is not relieved it is very difficult to get mucus up and out of those airways.

Removing the trigger and/or using drugs called **bronchodilators** are ways of treating bronchospasm. Ventolin (Albuterol) and Theophyline are common bronchodilators.

Color of Sputum: The color of sputum also gives important information about infection. Sputum normally should be clear or white. Yellow mucus means there is an infection and green mucus means there is not only an infection but there also is old "gunk" (or pus) that has been down there for a while. It usually is a sign of chronic infection. Blood in the sputum (which is called hemophysis), is important and should always be called in to the CF clinic if it is new and more than just a few streaks of blood in your sputum. This is usually a sign that the infection in your lungs has gotten somewhat worse, that it has actually bruised some of the blood vessels in your lung. Make sure you keep close track of how much blood you cough up. Occasional blood-tinged sputum can be common and is nothing to get real upset about. But large amounts of coughed up blood in the sputum can be more serious, even life threatening. Measure the amount by easy-to-understand amounts such as teaspoon(s),

tablespoon(s) and cup(s), rather than "small" amount or "average" amount, which can mean different things to different people. And when it comes to how much blood or sputum you are coughing up you want to be as specific as possible. With hemoptysis check with your CF HCP is you need to increase the amount of Vitamin K you are taking.

Good huffing and coughing is also an excellent way to treat your infection. Each time you cough out sputum or swallow it, you are eliminating some bacteria from your lungs. Make it a habit to cough regularly (like every time a commercial comes on TV - cough, or when the phone rings - cough, or every time you take a study break - cough). It will do wonders for your lungs. Try it!

Consistency of Sputum: Lastly, assess the consistency of your sputum. How thick or thin is it? You can get a good idea of this by coughing some sputum into a tissue and then opening and closing the tissue to see how easily the sputum moves. Over time, when you have colds, for example, you will be able to see how your sputum gets thicker and stickier. When it is like this, it is more difficult and more work to move it up and out of your lungs. Drinking lots of water (six to eight 8 oz glasses of water each day) is recommended and doing aerosols (such as saline, Mucomyst® and Pulmozyme®) are ways to thin your sputum. Many CF adults have found that drinking a large glass of water while using the Vest for their ACT makes it easier to cough up mucus.

Has your sputum changed? (yes/no)
If yes, how has it changed?
What is the amount of sputum you have coughed up in the past 24 hrs? __ none, __clears throat and swallows, ___tsp(s), ____, tbsp(s), ___cup(s).

What is the color of your sputum? __clear/white, __yellow, __green, __brown, __blood.

What is the consistency of your sputum? ___ thin, ___thick, ___thicker, ___very thick.

What is the status of your sputum?
1 (excellent/none), 2 (very good/scant amount), 3-4 (good/small amount), 5-6 (moderate/average amount), 7-8 (not very good/large amount), 9 (poor/very large amount), 10 (worst ever).

Circle your correct #: 1 2 3 4 5 6 7 8 9 10
Is this a change in the amount, color or consistency of your sputum? (yes/no) Should you do anything about these changes?

Suggestions: call your CF clinic/CF HCP, increase ACT, increase aerosols, take all prescribed antibiotics, increase fluids, check to see if you should take a sputum culture into the clinic.

• SHORTNESS OF BREATH (SOB)

Do you experience any shortness of breath (yes/no)?
Using the following scale, what is your status? 1 (none), 2 (small), 3-4 (mild), 5-6 (moderate), 7-8 (a lot), 9 (severe), 10 (worst ever).

Circle your correct #: 1 2 3 4 5 6 7 8 9 10

Is this a change from yesterday? (yes/no) If yes, is it better or worse? If worse, do you need to do anything about it?

Suggestions: call CF clinic/CF HCP, make sure you are using inhalers with a spacer or other meds (steroids, bronchodilators) as prescribed, increase frequency of ACT, increase aerosols, monitor shortness of breath.

• WHEEZE

Are you experiencing any wheezing? (yes/no)
Using the following scale, what is your status? 1 (none), 2 (small), 3-4 (mild), 5-6 (moderate), 7-8 (a lot), 9 (severe), 10 (worst ever).

Circle your correct #: 1 2 3 4 5 6 7 8 9 10

When do you experience this wheezing? __ at rest, __ light exercise, __ with hard exercise, __ coughing, __ cold air, __ smoke, __ other.

How often do you wheeze? __one to two times per day, __once every few hours, __once per hour, __several times per hour. Is this a change from yesterday? (yes/no) If yes, is it better or worse? If worse, do you need to do anything about it?

Suggestions: call the CF clinic/CF HCP, use inhalers with a spacer, use aerosols, use other meds (steroids or bronchodilators) as directed by your CF HCP or increase ACT.

Sidebar

Some individuals with CF also have asthma or reactive airway disease where there can be a lot of wheezing and shortness of breath. Most people with asthma have an **asthma action plan** for times when they wheeze more, have more shortness of breath and their peak flow meter reading has changed. If you have such a plan, make sure you are following it. If not, call the CF center/your CF HCP for advice on how you should manage this health situation.

** Mucomyst®* is another mucolytic or medicine that thins mucus. The CF Pulmonary Guidelines for chronic medicines has concluded that there is insufficient evidence for or against the routine use of Mucolmyst®. In this author's clinical experience in CF, Mucomyst® has been a helpful tool. MJM

• EXERCISE

What is your ability to be physically active (for example, run, do stairs, bike, dance, walk)? ❑ unlimited, ❑ average, ❑ a bit, ❑ rest a lot.

Using the following scale, what is your status? 1 (excellent), 2-3 (very good), 4-5 (good), 6-7 (fair), 8-9 (poor), 10 (worst ever).

Circle your correct #: 1 2 3 4 5 6 7 8 9 10

Is this a change? (yes/no). If yes, is it better or worse?

Sidebar

Exercise is a very important "therapy" in CF. The benefits are well known and have been proven over and over again in research studies. Just a few of the benefits of exercise are: it helps move mucus/sputum up and out of your lungs (just see how often you cough whenever you exercise), increases your vital capacity (one of your important lung measurements), strengthens the muscles you use to breathe, releases certain chemicals in your body that give you feelings of well being, feels good, improves your appetite. That's a lot of benefit! You should try to exercise 30 minutes a day, 3 times a week. Boy, would you feel great if you did that. When walking or exercising, make sure you drink plenty of fluids and replace the body salts lost with sweating (drink electrolyte drinks such as Gatorade, eat salty foods and liberally salt your food).

Many adults with CF use their workout, and their ability to do their workout, as a good way to monitor how they are doing. It could be their ability to increase their endurance over time (example: first game of hockey for the year serves as the baseline and from there you compete with yourself to raise the bar to get better and better). Or it could be the opposite and make you realize you are not able to do "today" what you were able to do yesterday or last week.

• CHEST PAIN

Do you have any chest pain? (yes/no) If yes, where is it?

Is it with inspiration (breathing in) or expiration (breathing out) or both? Using the following scale, how severe is the pain. 1 (no pain), 2 (small), 3-4 (mild pain), 5-6 (moderate pain), 7-8 (a lot of pain), 9 (severe pain), 10 (worst pain ever)?

Circle your correct #: 1 2 3 4 5 6 7 8 9 10

Is this new pain? (yes/no) Has the pain gotten worse/better/stayed the same? What makes the pain better or worse? Do you have limited chest movement on any side or do both sides move equally? (yes/no) If yes, how is it limited? Do you need to do anything about any changes in chest pain or movement?

Suggestions: call the CF clinic/CF HCP if chest pain is new and is affecting your ability to cough or do effective ACT or if pain is uncontrollable, monitor yourself for other changes, take Motrin® or Tylenol® for pain. Go to the emergency room immediately if your pain is intolerable and/or you have significant shortness of breath.

Sidebar

Any new onset of chest pain, which may be sudden, sharp or sticking pain that may radiate to the shoulder with difficult or labored breathing, may be a sign of a **pneumothorax** (collapsed lung). This is a medical emergency and you should go to an emergency room immediately. Most CF individuals who are in their late teens or older have developed overblown air sacs in their lungs. These are called "blebs." They are similar to an over-inflated balloon. These blebs can rupture and air can then enter the pleural cavity (the lining around the lung). The pressure may collapse the lung and displace the heart. This can be corrected first with a chest tube, which will re-expand the collapsed lung. Sometimes a chest tube is all that is needed to correct the ruptured area, but there are many times when surgery is needed. Having blebs does not mean they will rupture. Positive nutritional status and good body weight offer some good protection against blebs rupturing.

Before you leave the respiratory part of the body check, let's add one more piece to this self-care you are learning. Now that you are aware of the way your body can change and the signs it gives you when this happens, let's take it a step further. At your next CF check-up, establish some improvement goals for your Pulmonary Function Tests (PFTs) with your CF HCP. Then watch to see if you can find any changes that take place in your body check as you work to improve your PFTs. This will be interesting to watch – your own "research study"!

• GASTROINTESTINAL SYSTEM

Your gut, or gastrointestinal system, is very important in CF. It is now known that the better the nutritional status and body mass is for individuals with CF, the better they will be able to handle their lung disease. This cannot be emphasized enough! Most healthy adolescents and young adults with CF need approximately 3000 to 4000 calories each day and significantly more protein and fat than teens without CF. Your caloric needs will increase ebven more with lung infections and illness. You should spread your caloric intake between 3 meals and 2 or 3 snacks per day. Don't forget your digestive enzymes! If you are pancreatic insufficient you need digestive enzymes each time you eat or drink something nutritious. Remember, if you eat and don't take your enzymes, it is just like tossing the food down the toilet. You won't get the goodness or nutrients from that food for energy, growth and repair of your body. It is just a waste. Also

remember, foods that are high in fat will require more digestive enzymes for proper digestion and absorption. So, for example, instead of taking your usual 8 enzymes with a roast beef sandwich and chips, your may need around 10 to 12 enzymes with several pieces of pizza.

Sidebar

It also needs to be mentioned that as pancreatic-insufficient individuals with CF get older, their body's ability to handle simple sugars can become a problem. Scarring in the part of the pancreas that makes digestive enzymes can begin to affect the portion that makes insulin, usually in the teenage years and older. Many teens and young adults with CF become **"glucose intolerant,"** which means your pancreas makes the right amount of insulin for what you ate but it takes longer to do so. How you eat and what you eat become more important than ever.

Overall, you want to limit the amount of simple sugars you eat or drink by themselves. This means sugar-containing pop/soda is a great big NO! There is no nutritional value in pop/soda and it is a great big sugar load for your pancreas to handle. Sugar-free pop/soda is just fine. Eating a few candy bars is not a great choice either. But eating half of a peanut butter sandwich with a candy bar is better because you put some protein with it. Eating protein and fat with simple sugars will help your body handle the sugars better. Physical activity will also help.

For example, it is Halloween and you raid your little brother's candy bag and wolf down 3 candy bars with a Mountain Dew. Wow! That is a giant load for your pancreas's insulin making cells to cover. The best way to handle this situation is probably to jump on your bike and ride away from your angry brother! You should ride for about 20 minutes, which should give your little brother time to cool down and also help your body use that sugar load.

FYI… candy with nuts and chocolate have protein and fat in them, which makes them a better choice if you need a candy fix. Your CF dietitian is the expert who can help you learn how to make wise choices for meals and snacks.

• WEIGHT

Weigh yourself three times per week, on Monday, Wednesday and Saturday. NO MORE. Make sure you have a good scale. You can find these at any national bargain store for under $30.00. Get one - you will need it. Log your weight. Know what your weight goal is. Your HCP and/or CF dietitian should figure this out for you and how much weight you need to gain each week/month to achieve this goal. Work with the CF dietitian to figure out how many calories you need each day to gain weight and then how to get these calories in each day (meals and snacks).

What is your weight? _____ lbs. Is it the same, increased or decreased from your last weigh in? Are you meeting your weight goal? (yes/no) If no, why not? What needs to be done about it?

Suggestions: Call your CF dietitian for ideas on how to increase your calories, increase calories by adding a high-calorie snack right before bedtime, consider supplements, consider adjusting digestive enzymes, monitor the situation or add high-calorie shakes during the day. Keep a record of what you have eaten for several days and have the CF dietitian evaluate it for calories and ideas on how to improve it. Also, if your weight is a major problem consider gastrostomy or naso-gastric feeds.

Sidebar

A **gastrostomy** is a tube surgically placed in the stomach for receiving calories and nutrients when an individual is unable to eat enough food to maintain or gain weight for a healthy body. In CF, a gastrostomy is usually used to receive night-time feeds. This is when a liquid supplement drips into the gastrostomy (which is in the stomach) while the individual sleeps. The feeding is usually stopped a few hours before the individual gets up in the morning. This usually helps the individual tolerate their morning ACT better as well as eat a better breakfast since the stomach is not full. Night-time gastrostomy feedings can give an individual up to 800-1000 additional calories per day.

A gastrostomy can also be used for "bolus feed." This is when a certain amount of liquid supplement is infused over 20-30 minutes during the day while the individual is awake. This is another way of boosting caloric intake during the day. If one is pancreatic insufficient, they will require digestive enzymes with the gastostomy feeding. Having a gastrostomy does not mean you don't have to eat food. The same amount of caloric intake is usually required with the gastrstomy feedings serving as bonus calories to help one gain weight. Your CF HCP and CF dietitian can discuss a gastrostomy in more detail.

Naso-gastric feeds are similar to gastrostomy feedings except there is no tube surgically placed in the stomach. Instead, a temporary, thin flexible tube or catheter is passed through the nose down to the stomach. This tube is then used to drip a liquid supplement into the stomach usually at nighttime when the individual sleeps. This tube is usually removed in the morning once the feeding is complete. This is the same concept as a gastrostomy except the tube is temporary.

Because weight gain and body mass are so important in CF, special attention and emphasis must be given to it. You need to approach weight gain scientifically. That is, there is a set weight goal that needs to be established and then

broken down into weekly goals. This weekly goal should usually be one pound. You need to work with the dietitian on how many calories you need each day to achieve your weight goal and how that looks in meals and snacks for the day. So, for example, you should know that if you need to gain 10 pounds, in order to gain one pound per week, you will need 4100 calories per day. You know your breakfast is 1600 calories with your high-calorie supplement drink, bagel with cream cheese and jelly and cereal with whole milk. Lunch should be 1400 calories with a roast beef sandwich with mayonaise, chips, dessert and whole milk. Dinner should be 1300 calories with pork chop, mashed potatoes and gravy, broccoli with cheese sauce and chocolate whole milk to drink. This totals 4300 calories for the day. You are 200 calories above your needed amount so if you continue to eat like this, you should achieve your weight goal easily. (The calories mentioned here are not accurate and only serve as an example.)

If you are not gaining weight and you know you are taking in the correct amount of calories, you need to look at your digestive enzymes to see if they are correct and also check your blood glucose level. If you are not taking in the appropriate amount of calories and don't think you can, you need to be honest with yourself and think about a gastrostomy tube (g-tube). Putting on the weight is important; the g-tube is just a means to an end. Once the weight is on, you might not need it anymore. Without the weight, you will never get strong or fully develop the way you are supposed to.

• APPETITE

How is your appetite? Use the following scale: 1-2 (excellent), 3-4 (good), 5-6 (variable), 7-8 (not hungry), 9 (poor), 10 (worst ever).

Circle the correct #: 1 2 3 4 5 6 7 8 9 10

Is this a change? (Yes/no) Is it better or worse?
Do you have any nausea and/or vomiting? (yes/no)

If yes, how much and how often?
Is it better or worse?

What makes the nausea and/or vomiting better or worse?
Do you have any heartburn or reflux? (yes/no)

If yes, using the following scale, what is the status? 1 (none), 2 (small), 3-4 (mild), 5-6 (moderate), 7-8 (a lot), 9 (severe), 10 (worst ever).

Circle correct #: 1 2 3 4 5 6 7 8 9 10

Is this better/worse/the same as yesterday?
What can you do about it?

Suggestions: call your CF dietitian or HCP with nausea and/or vomiting and reflux concerns; with nausea and vomiting be aware of dehydration and drink plenty of fluids; consider using supplements; push high-calorie drinks, meals and snacks; consider tube feeding; monitor situation.

• ABDOMEN

Do you have any abdominal pain? (yes/no) If yes, where is it and how severe is it?

Circle the correct #: 1 2 3 4 5 6 7 8 9 10

Is this new pain? (yes/no) How long have you had this apin? Is the pain better/worse/the same as yesterday? What makes the pain better or worse? Are you pooping (bowel movement, stooling) regularly? (yes/no) If no, when was your last BM/stool? Do you have any abdominal bloating? (yes/no) Gassiness? (yes/no) Do you need to do anything about your abdominal complaint?

Suggestions: call the CF clinic/CF HCP, make sure you are taking your enzymes, call CF HCP to discuss adjusting your enzymes and/or need for medications Mucomyst®, Golytely®, or Miralax®, increase fluids.

Sidebar

It is common for individuals with CF to develop a partial bowel obstruction. This is called DIOS (distal ileus obstruction syndrome) or meconium ileus equivalent and, although it can happen anytime, it is most common during the summer months when it is hot outside. Dehydration and not taking digestive enzymes properly usually cause thick mucus in the bowel to mix with undigested food and then stool and cause a blockage, usually in the right lower quadrant of the abdomen. Clues of DIOS usually are that you haven't been taking your enzymes as you should and you are almost always constipated. Any time you go two days without having a bowel movement/stool, call the CF clinic! This is the tip off! You may be headed toward DIOS and should talk to your CF HCP about needing medications such as Mucomyst®, Golytely® or Miralax® to get things moving (to help you have a BM)! A third sign is abdominal pain. It is usually in the right lower side but could also be by your belly button. As the obstruction/blockage worsens the pain will worsen and eventually you will begin to vomit. This is the fourth sign. If you are now vomiting and have waited this long to do something, go to the emergency room immediately! THIS IS SERIOUS!! DIOS is easy to prevent by taking your enzymes and drinking plenty of water, especially during hot weather.

• STOOLS

You should usually have one to two average-sized bowel movements (BM or stools) per day with the majority normal-looking (round and soft) and they should sink in the toilet water, not float. An occasional greasy/fatty stool that floats in the toilet water is okay as long as it is only occasional. The reason it floats is it contains undigested fat. It would be better if your body absorbed the fat calories and not the toilet. If you have quite a few "greasy floaters" and/or more than two or three average-sized stools per day you need to talk to your CF HCP about increasing or changing your digestive enzymes until you have one to two stools per day and the majority sinks in the toilet water.

The dreaded **"rectal prolapse"** is a protrusion of the rectum through the anus outside the body. This is almost always due to **NOT** taking (or not taking enough) digestive enzymes. The poor rectum cannot handle passing all that bulky, heavy stool full of undigested protein, fat and carbohydrates. It gets tired and just falls out! Now that's what I call having a bad day!! Passing a lot of gas or wind ("farting") is another sign you are not taking enough digestive enzymes. So, do the people around you a big favor and either take your enzymes like you are supposed to or ask your CF HCP to increase your dose.

"Snake-like" stools look just like a snake - long and thin. Anytime you have this type of stool, be careful. This often precedes a partial bowel obstruction. If you haven't been taking your enzymes properly, you should start. If your stools are mucousy, ask your CF HCP if you should begin taking Mucomyst®. If you haven't been drinking enough water, drink six to eight 8 oz glasses of water each day. These efforts should help prevent DIOS. If you are having abdominal pain, call your CF clinic right away.

How many stools did you have yesterday
(___1 ___2,___3,____4, ___more)?
Are they usually ___normal, ____greasy/fatty, ___floating, ___gassy, ___diarrhea, ___constipation, ___snake-like?

Any rectal prolapse? (yes/no)
If your stools are not what they should be, what do you need to do?

Suggestions: call your CF clinic/CF HCP to discuss changing enzymes or adjusting dosage, discuss the need for Mucomyst®, Miralax®, Golytely®, take your enzymes regularly, adjust enzymes better when eating fatty foods, drink plenty of water.

• EXTREMITIES

Any swelling of ankles, feet, fingers or hands? (yes/no) If yes, where is the swelling and how long have you had it? Is the swelling better/worse/the same? Do you bruise easily? If yes, are you taking vitamin K regularly? Is this a change? (yes/no) Is the bruising better or worse?

Using the following scale, what is the status of swelling or bruising in your extremities? 1 (none), 2 (small), 3-4 (mild), 5-6 (moderate), 7-8 (a lot), 9 (severe), 10 (worst ever).

Circle your correct #: 1 2 3 4 5 6 7 8 9 10

Do you need to do anything about it?

Suggestions: call your CF clinic/CF HCP, take your medications as prescribed, ask your CF HCP if you should increase vitamin K dose, monitor the situation.

• PAIN

Do you have any pain in your head, chest, stomach, joints, abdomen or any other place? Is this new pain? (yes/no)

Using the following scale, what is your status?

Circle your correct #: 1 2 3 4 5 6 7 8 9 10

Is the pain better or worse than yesterday? What makes it better or worse? What do you need to do about it?

Suggestions: use ibuprofen (Motrin®) or acetaminophen (Tylenol®) as needed, call CF HCP if pain is new, persistent or not controlled with over-the-counter medicines. Check with your CF HCP regarding how much ibuprofen (Motrin®) or acetaminophen (Tylenol®) is safe to take every 24 hours. These medications can be toxic when taken in large quantities.

• OUTLOOK

Overall, how is your mood/outlook? ___Are you cheerful, __content, __happy, __serious, __variable, ___sad, ___tearful, ___irritable, ___angry, ___frustrated, ___whiny or ___depressed?

Is this a change? (yes/no). If yes, is it better or worse? If worse, what do you need to do about it?

Suggestions: talk with one of your support people, call the CF clinic, or talk with a psychologist or CF social worker.

• OTHER

This is for you to personalize. If you are diabetic, put your glucose checks here. If you have a port-a-cath, Hickman or PIC line, here is where you would put your line care, checking to see if it is red, tender, any swelling, any discharge or drainage, if the line has a good blood return and if it infuses well. If you have a gastrostomy or a MIC-KEY®, here is where you would check that site. Is there any redness, tenderness, discharge, drainage or foul odor?

Now that you have gone through this body check and thought about each area of your body specifically, how would you rate your overall post-body-check status?

How do you rate yourself from 1 to 10 using this scale: __1 (excellent), __5 (fair), __10 (worst ever)?

Circle your correct #: 1 2 3 4 5 6 7 8 9 10

How does this number compare with the number you gave yourself at the beginning of your body check? Hopefully they are the same or close. If not, spend some time checking out why they were so different and what you learned doing your body check to change your status. Each day your body check will get easier and easier to do.

The important thing to keep in mind is that your body is trying to tell you things about how it is functioning. We need to learn to listen and look for the right clues to figure out what is going on. And since it is your body, you are going to be the best person to know what is right or wrong, fixed or broken or just not quite right.

Congratulations on completing your BODY CHECK!! You are becoming an expert in self-care.

Table III in the Resource Section (on page 45) has the Body Check Log that you can use each time you do your Body Check until you get good enough to do it in your head. Photocopy it so you have multiple copies to use and remember to take a completed Body Check Log to your next clinic visit.

As mentioned earlier, the team leader, which is you, has four different jobs. The **FIRST** one, assessment and decision-making for your health, was covered in the Body Check.

The **SECOND** responsibility is making return appointments with the CF clinic and scheduling any tests or procedures recommended by your CF HCP, as well as scheduling appointments with other doctors, psychologists or other professionals as needed.

Your **THIRD** responsibility is managing your supply of home medications and medical supplies, which can be an enormous job. The critical point of this job is to never run out of medications and to come up with a system that will make it manageable and not drive you crazy. There are several good systems that work well. First, if your CF clinic is in a large medical center with a pharmacy, it is easy to set up a system with them. Every time you go to the clinic, you refill your meds at that pharmacy. Make sure to call the day before so they will be ready for you. Then, during your CF clinic visit, you can review your meds with your CF HCP and the clinic can call any changes directly into the pharmacy. Then, your meds will be waiting for you after your clinic visit. If your insurance company will only pay for one month of medications at a time, arrange for the pharmacy to mail your medications monthly until your next clinic visit. Make sure you use your calendar to keep track of when your med supplies run out and when your order from the pharmacy is due to arrive. You should call the pharmacy at least 5 business days before your order is due to remind them so nothing is left to chance.

The second system you can use is to get your medications from the Cystic Fibrosis Services Pharmacy, a subsidiary of CFF. Your CF clinic will send them your prescriptions and they will mail your meds to you. You arrange each month to have your refills mailed to you the same way you would with the CF center pharmacy. A good reason for using either of these systems is that they buy medications in such a large quantity, they can sell them at a less expensive cost to you and the drugs you need will always be available. For most local pharmacies, this is not the case.

When it comes to your medical supplies, if you need any, a system like the above examples will also work. Most of the medical supplies you need will be available through your CF center pharmacy and you can order them along with your meds. It is a good idea to always have one week of extra medical supplies on hand for emergencies, a last minute trip or whatever. You just don't want to ever run out and this way you are always covered.

Your **FOURTH** and final responsibility as team leader has to do with health insurance. This can be a simple or very challenging job but it is always a critical one since healthcare is very expensive and CF is a costly disease. The first part of this responsibility is to know and understand your (or your parents) health insurance policy. This means understanding what is a covered (paid for) medical expense, what is not covered and what is the coverage (what percent of the cost is covered and what is your responsibility). You also need to be aware of any specific rules/regulations they may have on specific coverage. Examples are using generic medications, getting prior authorization for certain tests, medication, hospitalizations etc. Every insurance policy is different so don't make any assumptions or guess about your coverage – it can be a costly mistake. Health insurance today can be very complex with lots of rules and it is your job to know what they are. Part of growing up and becoming an adult is learning to be knowledgeable about your health insurance coverage.

It is important to go through all medical bills and explanations of health care benefit forms from your insurance company for accuracy and possible mistakes. Errors happen and they can cost you money if undetected. Pick one day a month to go through these items so you stay up to date. It is best to develop a filing system that works for you to keep all your health insurance papers in order.

It will make things easier for you if you can get a **case manager** assigned to you by your insurance company. This can then be one person for you to call with questions, concerns or to help advocate your position. If you don't have a case manager already, ask for one.

A major consideration for you will be health insurance coverage once you are no longer on your parent's policy. Policies differ but many cover full-time students between 18 years to 21 or 25 years of age, therefore most young adults with CF go to college following high school graduation to continue coverage under there parents policy.

Because you are chronically ill you are considered uninsurable or unable to obtain health insurance on you own. Most chronically ill individuals obtain health insurance through their employer who offers health insurance as a benefit. Before accepting any job offer, make sure you thoroughly check out the health insurance coverage they offer. Check to see if you would be able to continue to receive care at the CF center, what coverage you would have for medications (oral & IV), out-patient and in-patient care as well as home care.

Your best resource when is comes to health insurance is your CF social worker. He/She can be a big help with problem solving as well as assessing health insurance coverage and whether it is right for you.

Professional CF-Self Care Team

Now that we have covered your responsibilities as team leader, let's move on to setting up your CF self-care teams. You will have three self-care teams that will be available to help you successfully manage your CF using self-care.

The first self-care team is your **Professional CF Self-Care Team**. This is your CF clinic team. Each CF clinic is different across the country with some having only one physician, while others may have several physicians and nurse practitioners. If you have a choice, you need to choose a "Primary CF Health Care Professional" (CF HCP). This person will lead your professional CF team and will see you each time you go to clinic. If you have not begun being seen alone in clinic, without

your parents, now is a good time to start. This gives you a good opportunity to develop a relationship with your CF HCP as well as ask questions you may not want your parents to hear.

Also on your professional CF team are the other CF team members, as well as clinic and hospital staff. These are all the professionals who are available to help you meet your goals of self-care and point you in the right direction.

Personal CF Self-Care Team

The second team is your **Personal CF Self-Care Team**. This includes you and your support people. Here we have to get very specific. You need to identify support people who will be available to help you with your CF cares when you need it or should you need it. You may need your mom to clean your equipment every day as well as to make your high-calorie shakes three times a day except on weekends, when your sister has agreed to do this for you.

Another example is before finals week, you and your parents sit down to discuss who will take responsibility for which CF cares. This strategy will help you stay well and get your CF cares done while you put in long hours studying. The plan you develop might have your father responsible for having aerosol solutions ready to use 2 or 3 times a day with your vest, mom responsible for cleaning your equipment daily and making your high-calorie shakes three times a day and your sister being extra nice and agreeing to do your chores that week! What a gal!

Some of your support people will be available for help with actual CF cares while others will be there for emotional support. Both kinds are needed for long-term and life-long success.

PRN CF Self-Care Team

The last team to set up is your **PRN CF Self-Care Team**. PRN means "as needed" in Latin. This team will only be needed in unusual or emergency situations. An example is your mother broke her arm and you have no other family members living in town. Your school nurse is on your PRN CF team and therefore could take over mixing your aerosol solution while your best friend's mom (also a PRN team member) cleans your equipment every night. You are able to do all your other CF cares independently. Your best friend's dad could also be on your PRN team since it is a good idea to have an extra person for good measure.

You never know when someone might get sick or go out of town. Better to be over-prepared than up a creek without a paddle! Many CF centers also have Home Health Services that can send a nurse and respiratory therapist to your home to help with home cares when needed. These services may be covered by health insurance. A nurse from a home health service will be especially helpful if you should ever need intravenous (IV) antibiotics at home.

Remember, you are the leader of your CF Self-Care teams. Since it is your body we are talking about, you are the boss. This is a lot of responsibility but there are a lot of people to help you make decisions. The people at the CF clinic are experts and are there to help you. It is important to develop a good relationship with your primary CF HCP. Feel free to call the CF clinic anytime you have questions that can't wait until your next clinic visit. It is a good idea to have a notebook just for your CF stuff with a section for information, clinic notes, questions, etc. Remember to write your questions in your notebook so you can ask them the next time you go to clinic.

C. Recognize Levels of Self-Care Change

Self-care is all about taking good care of yourself and doing what it takes to keep yourself healthy. That is the goal. There will be times when you will need more help from your teams to achieve your goals and there will be times when less help will be needed. This is called "different levels of self-care." Different factors influence one's level of self-care. Family, physical, psychological and social factors can impact how much or how little self-care one can successfully handle.

Family

The first factor is family. Many parents embrace self-care early in life, encouraging their child with CF to assume responsibility for age-appropriate CF cares. Other parents do the opposite, keeping total control of all CF cares and over-protecting their child and teen with CF. Other families fall somewhere in the middle. In most families, getting CF cares accomplished is a battle. Hopefully, self-care can help you and your family eliminate many of these battles.

Physical Issues

The second factor has to do with physical issues. When you are run down or sick, CF can often rob you of a good portion of your physical strength. When energy levels and physical strength are low, performing CF cares can be difficult. At times like these, asking for help is a smart thing to do. This way you are able to conserve your energy and hopefully prevent getting really ill.

Emotional Health

Emotional health affects your energy levels. Being a teenager can be very difficult. This is a time when it is common to feel unsure of yourself and to question who you are. Feeling unattractive, unpopular or having low self-esteem is fairly typical for teens, especially teens with chronic illness. When this is the case and you feel low in emotional energy, you may not have the necessary energy needed for self-care. It is important that you realize this and let your team members know that you need more help in getting your CF cares done.

Recognize your limitations and then do something about them. This is a critical point in self-care. You need to learn what you can and can't handle and then plan accordingly.

Depression

Nothing robs one of energy more than depression. It is critical to recognize the signs and talk to your parents and/or CF HCP about getting some help. **Common signs of depression include poor appetite, weight loss, change in sleep habits (too much or too little), little interest in things and falling grades.** Admitting that you feel depressed is a very difficult thing to do but is one of the smartest. Depression is a common occurrence in chronically ill adolescents and if untreated, depression can become severe and, in some cases, can lead to suicide. It is very important to talk to someone before your feelings get too low. This can be one of your team members, your CF HCP or even a psychologist. This is a very healthy and wise thing to do. Don't be afraid of what others might think; your health is what is most important.

It Won't Happen to Me

"It won't happen to me" is an age-old fantasy of most adolescents. Don't kid yourself, it can happen to you. It can, and it will if you don't take care of yourself! Your self-care is directly related to you future health. What you don't do today can hurt you tomorrow, even if you don't think it will. In CF there is a high price to pay when you don't take care of yourself: YOUR LUNGS AND THEN YOUR LIFE.

Social Life

The last factor to consider is your social life. Finals and homecoming week are both typical high-risk social situations. Studying and extracurricular activities with late nights sometimes require energy above and beyond the normal energy you need to do your everyday tasks plus your CF cares. Often, CF cares fall to the bottom of your priority list and get left for another day.

You might think that missing your CF cares for a day or two is no big deal. Well, think again! You really need to think again, literally! We all need to cheat once in a while; only a robot does everything all the time. However, you need to be smart about how and when you cheat! First, if you are sick, DON'T CHEAT! Only negative things can happen. If you are feeling well and your body check supports this, go ahead and fudge on your CF cares for no more than a day or two. And to be REALLY SMART, make sure you do a really good job with your CF cares the next day. Remember, CF is sneaky! So, be smart and plan ahead. Arrange for help with CF cares if you need it so you stay well and healthy to make it to all of your social events feeling great and having a blast!

D. Who Is Going to Do What?

It is time to sit down with your parents and establish who is going to do what CF cares and when. Letting go and having you take responsibility for your CF cares can be very stressful, even scary, for your parents. They have spent your whole life working hard to keep you healthy and prevent serious CF-related complications from occurring. Turning that over to you is not going to be easy. Doing a really good job managing your CF cares will help decrease some of their fears and begin to win their confidence and trust in your abilities.

Setting up a Schedule

If you don't already have a daily schedule, now is the time to start one. If you do, you need to add your CF cares. Starting from scratch, the first step is to list all your CF cares and the time of day they are needed. Set this aside. Next, begin your schedule with the start of your day and list your daily activities from the time you get up in the morning to the time you go to bed at night, including school, time with friends and studying. Try to be as complete as possible so you get an accurate picture of your life. Now take your CF cares list and plug them into your daily schedule. The following example is a list of a teen's CF self-care activities.

This is a list of daily self-care activities for a typical teen with CF.

- Prepare morning aerosols
- Morning ACT: (example uses Vest for ACT)
  Albuterol/saline neb (done at beginning of Vest treatment)
  Pulmozyme® neb (during Vest treatment)
  Tobi® neb (done at the end of Vest treatment)
  Coughing/huffing throughout ACT
  Specifics of ACT vary between CF centers.
  Follow what your CF HCP has prescribed for you.
- Weight check (Monday, Wednesday, Saturday)
- Body check
- Morning meds
- Digestive enzymes with breakfast
- Noon meds
- Digestive enzymes with lunch
- Cleaning aerosol equipment
- Exercise for 30 minutes (Mon, Wed, Sat)
- Evening meds
- Digestive enzymes with dinner
- Prepare evening aerosols
- Evening ACT: (example uses Vest for ACT)
  Albuterol/saline neb (during Vest treatment)
  Tobi® neb (at end of Vest treatment)
  Coughing/huffing throughout ACT
- Estimate calorie intake for day
- High-calorie shake with digestive enzymes before bed

You will probably need two different schedules, one for the weekdays when you have school and one for the weekends when you don't (and can sleep in). Look at your schedules and discuss with your parents what seems realistic for you to do and what doesn't. Remember to be very clear about who will do what and when.

For example, mom will clean all equipment Monday through Friday but you will do it on the weekend. Plan for success, not failure. Be a team and come up with a winning combination that will keep you well and help you become more independent over time. Be careful not to take on too much and then not be able to handle it, miss therapies and maybe get sick. Let's be smart about self-care.

The following chart is an example of what a daily self-care schedule might look like with cares broken down between a teen with CF and his or her parents. Notice how they differ on the weekend.

Example of a CF Self-Care Schedule

	Monday	Tuesday	Wednesday	Thursday	Friday	Saturday	Sunday
5 a.m.							
6 a.m.	• Get up • ACT with nebs • Body Check					Sleep in	Sleep in
7 a.m.	• Shower • Weight Check • Morning Meds	• Shower • Morning Meds	• Shower • Weight Check • Morning Meds	• Shower • Morning Meds	• Shower • Morning Meds		
8 a.m.	• Breakfast with Enzymes • School bus (8:30)						
9 a.m.	• School						
10 a.m.						• Get up • ACT with nebs • Body Check • Weight Check	• Get up • ACT with nebs • Body Check • Snack w/Enzymes
11 a.m.							
12 Noon	• Lunch with Enzymes						
1 p.m.							
2 p.m.							
3 p.m.	• Home from School						• Big Snack with Enzymes
4 p.m.	• Ride bike (30 min.) • Snack with Enzymes	• Snack with Enzymes	• Ride bike (30 min.) • Snack with Enzymes	• Snack with Enzymes	• Snack with Enzymes	• Ride bike (30 min.) or walk	
5 p.m.					• ACT with nebs • Evening Meds • Estimate Calories for Day	• ACT with nebs • Evening Meds • Estimate Calories for Day	
6 p.m.	• Dinner with Enzymes • Evening Meds				• Dinner with Enzymes	• Date for dinner & movie (take enzymnes with you)	• Dinner with Enzymes • Evening Meds
7 p.m.	• Homework				• Football game & dance (take enzymnes with you)		• Homework
8 p.m.							
9 p.m.	• ACT with nebs • Estimate Calories for Day						• ACT with nebs • Estimate Calories for Day
10 p.m.	• High Claorie Shake with Enzymes • Bed						• High Claorie Shake with Enzymes • Bed
11 p.m.							
12 p.m.							

This schedule is the same Monday through Friday except for weight checks, which are Monday, Wednesday and Saturday. Schedule is different on Saturday and Sunday. ACT includes aerosols with treatment vest and coughing/huffing.

CF Self-Care Contract

Now that you have decided who will do what CF cares and when, it is usually helpful to have a contract signed between you and your parents. This ensures everyone understands who is responsible for what and when, and will do their best to stick to it. It also makes it a big deal and tougher to break. Using rewards or reinforcers for following through with your contract is highly recommended. Let's face it: CF is hard work! Anytime you can earn a reward that says "good job" or "well done" is important. It is these rewards that can help you get through those days when you are really dragging. Thinking about a big juicy steak from your favorite restaurant (your chosen reward) can give you the push you need to get up off the couch and start your treatment. You should come up with a potential list of rewards that work for you and then sit down with your parents to discuss which ones they can agree to. Monetary rewards should be discouraged and activities that bring families together should be encouraged.

The following page is an example of what your Self-Care Contract might look like. There is a blank contract in the Resource Section, Table V, for you to copy and complete with your family. Once your contract is complete you may want to share this with your Professional CF self-care team.

Example of a CF Self-Care Contract

I, _____ (name), in an effort to become more responsible for my health, willingly agree to this self-care contract.

I will be responsible for:
(list all CF cares you will do).

For Example:

1. All ACT (doing all my aerosols and vest) twice per day
2. Take all medicines
3. Take all enzymes (especially with snacks)
4. Exercise three times per week
5. Gain one pound per week
6. Drink a high-calorie shake daily

Parents will be responsible for:
(list all CF cares mom/dad will do)

For Example:

1. Prepare all aerosols (including Albuterol®/Saline Pulmozyme® & Tobi®) two times per day
2. Sit with you during ACT (dad will do)
3. Clean aerosol equipment daily (mom will do)
4. Make high-calorie shake daily (mom will do)
5. Ride bike or go to health club with you at least three times per week (mom or dad will do)

Reinforcers/Rewards:

For Example:

1. Choose dinner for family movie night
2. Use the car one day on the weekend
3. Have your chores done for you for one week
4. Get a 20 minute back rub or massage
5. Go on a weekend camping trip with friends
6. Two tickets to movies of your choice and money after for pizza
7. Dad takes me duck hunting
8. Choose movie and dinner for family movie night

For Example:

1. Do all CF cares for five days
2. Fullfill your contract for one week (CF cares & gain one pound)
3. Fulfill your contract for two weeks
4. Do all CF cares for three days
5. Fulfill your contract for one month
6. Fulfill contract for 2 weeks
7. Do all CF cares for two weeks
8. Do all CF cares for one week

Signed

Teen_____ Date:_____

Parent(s)_____ Date:_____

Hey, Does Anything Need to Change?

Now that you have all the pieces in place, you are set to go. Give these ideas a test run and see how things go. After a few weeks or a month, sit down with your parents and review how it is going. What works? What doesn't? Even good plans need some adjusting now and then. Your self-care plan may also need to change. Take a close look at all aspects of it and see how it is working for you. (That is a favorite Dr. Phil line, "How's that working for you?") Don't be afraid to make changes. Life is a work in progress. You change, so your self-care plan should also change. Just keep on top of things and be one step ahead.

III. The Balancing Act

A. Putting It All Together

Have you ever gone to your CF clinic appointment feeling good about the way you have been managing your health only to find out that you are not doing as well as you thought you were and have lost weight? Do your CF HCP and mother talk about you as if you were still a kid? If so, this doesn't feel very good. You probably feel frustrated, unheard and even angry. Many times you leave with a new medical plan you had no part in developing.

This is a pretty typical scene in the lives of teenagers with CF. One of the biggest challenges is developing a medical plan that works with your real life. Fortunately, there are ways that you can learn to work with your medical team and parents to develop a balanced lifestyle that will lead to success. Here are some tips for putting it all together.

Don't sit there with a blank look on your face.

- Start talking and asking questions. If you want to be involved in medical decsions, let yourself be heard. Take the lead in answering your CF HCP's questions rather than letting your parents answer for you.

- Educate yourself. You are older now and need to understand more about CF. Understanding CF will allow you to feel more in control. Good knowledge will lead to better decision-making. Your clinic will have educational materials available to you. Use them. Also, use the internet. A list of good CF websites is in the resource section on page 51.

Taking care of yourself should be a part of your everyday life.

- Just like homework, it's easier to follow your medical plan if you see it as just another thing to do every day. You may not like doing it, but it helps you do better in the long run. Make taking care of yourself a part of your routine, like doing your homework, or brushing your teeth.

Find a schedule that works.

- Review your self-care schedule to see if it is working for you. If it is not working, then change it.

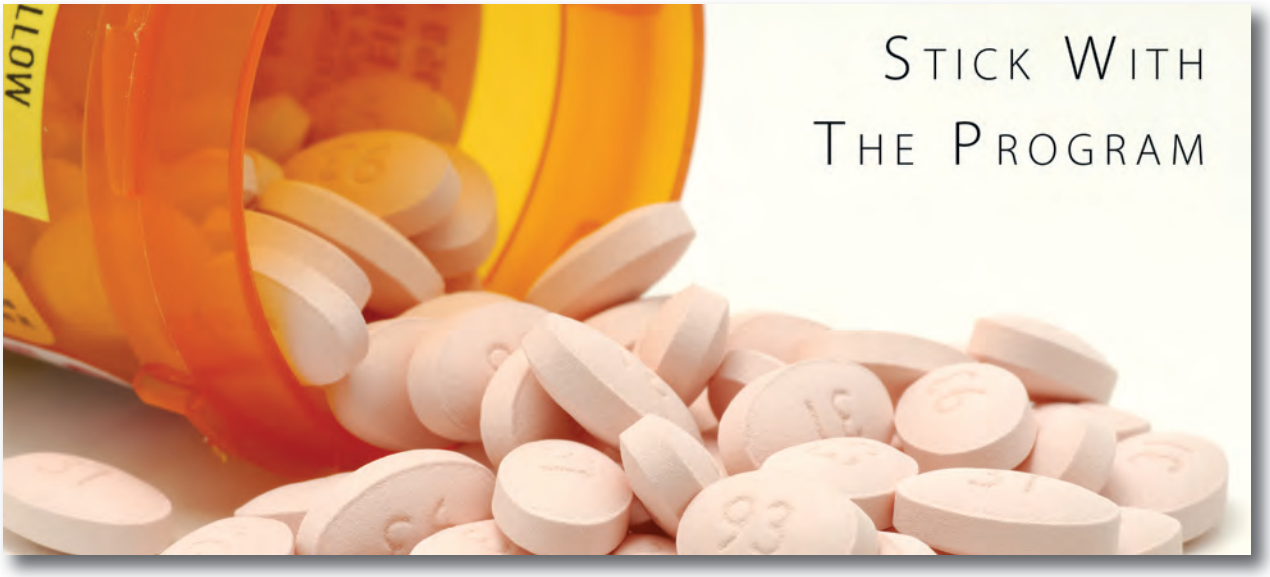
Develop a plan to tackle the hard stuff.

- Choose a specific behavior to change. There are probably a few areas of your self-care that are difficult for you to do. For many teens, this includes diet and exercise. The best thing to do is to choose one area to focus on at a time.

- Start out small. If your goal is to exercise more, begin by exercising three times per week and not every day. As you meet your goals, develop new ones that are more challenging.

- Reward yourself. As discussed in the Self-Care chapter, using rewards is important. This is hard work and you deserve a treat! Again, work with your parents to come up with good ideasfor rewards that will work for you.

- Keep going. Once you have met your goal, don't slack off! To keep this from



happening, set a long-term goal with a special reward for yourself. For instance, exercising three times a week for three months and rewarding yourself with a special weekend event.

Show your parents you can do it.

- Take responsibility. The best way to show your parents you are no longer a kid is to take responsibility for part of your medical care. Talk to your parents about things that you thinkyou can handle on your own. Remember, it might be difficult for them to let go.

- As mentioned in the Self-Care chapter, develop a self-care contract. This should spell out clearly who does what and when.

- Use your self-care schedule to stay on track.

- Remember it's all about balance and changing your level of self-care as needed. Sometimes you can do more and sometimes you will do less. Be smart and use your CF Self-Care teams as needed.

Know when and where to get help.

- Hard times: All teens have hard times, especially those with chronic illness. Pay attention to how you are feeling both physically and emotionally. Is your body trying to tell you something? You will need to make adjustments according to how you feel that day and what else is happening in your life. Remember to adjust your CF Self-Care teams as you need them. Remember they are there to help you so use them.

Don't give up!

- It is important to talk with your parents and CF professional team to develop a plan that fits into your life. It may take some work and compromise by everyone, but eventually you will be able to find the right balance for you.

B. Adherence

Most people with CF are expected to take many different medications and treatments. And, as one gets older, the list usually grows! Many of these, such as airway clearance or aerosol treatments, can be time consuming. Some medications just taste bad, while others can cause side-effects such as nausea. Although the benefits of some medications may be clear (such as pancreatic enzymes), you might wonder why you need to take others.

If you are not convinced that a particular medication is making a difference, discuss it with your HCP. You should know what medications and treatments are prescribed for you and why. Be smart with your medications and take them as prescribed. Discuss with your CF HCP how to prioritize your treatments so on really hectic days you understand what must be done.

Finding time to fit in all of your meds and treatments can be a problem, especially as most people get busier as they get older. Are there shortcuts you can take? Are you taking your medicines at different times of the day when it might be easier to take them at the same time? Find out which meds can be taken together and which cannot.

Talk to the CF team if your CF care seems to be taking up too much time. There may be some aspects of the treatment regimen that can be changed. CF cares can "eat" a good portion of each day. Use the following tips to help you fit CF care into your daily life.

Make it a priority. Life should not be focused on CF. If you have significant CF complications, life will not be as enjoyable as when your CF is better controlled. Therefore, work on preventing complications by doing your ACT regularly. Most CF adults find that taking care of themselves (doing their CF cares) is the best way to stay healthy and get the most out of life.

Develop routines. Most people find that developing treatment routines is the only way to turn something into a habit. It can be helpful to use a friend or parent to remind us when we are trying to develop new habits, like exercising regularly. Activities are more enjoyable when done with other people. Instead of jogging alone, find a regular jogging partner.

Find fun things to do

Find enjoyable or distracting things to do while you're doing your treatments, such as listening to music, watching TV, using the computer or playing a game. You can contract with a friend to be with you and chit-chat during ACT. Remember to use rewards!

Most people find ways to fit CF into their adult lives so that they are in charge rather than CF. However, doing your treatments day-in and day-out can sometimes be tougher to do than at other times. Everyone takes time out from treatments occasionally, but don't make a habit of it. Sometimes, especially when you are feeling too busy or particularly low, you might feel that you just can't be bothered with it anymore. This can be normal. However, it is good to talk to your family, friends and others you've identified as your support people. Your Professional CF Self-Care team can help you connect with other adolescents and young adults with CF. Learning how to ask for help can be hard, but when the going gets tough, doing it on your own is no fun. We all need a little help from our friends.

C. Pitfalls to Be Aware Of

Life as a teenager can be rocky at times - juggling school, family, friends, work and activities. For teens with CF, there are even more things to juggle, like health, ACT, diet, exercise, medications and enzymes. That is a lot for anyone to think about. However, you should know that most teens with CF are well adjusted. There are going to be times when you may feel that you are juggling too much and may want to drop everything. You shouldn't feel helpless during these times. Here are some common areas that are stressful for teens with CF and some tips about what you can do if you experience them.

Feeling Different

Sometimes you just want to be like everyone else. It may seem unfair that you have to struggle with things that other teens don't. You may also start to feel like everyone is noticing that you look different (don't all teens!). But here's a secret: Some teens are so worried about how they look and act they don't notice much about other people. It's part of being a teenager! Therefore, the best advice is to try to accept and be comfortable with yourself just the way that other teens don't. You may also start to feel like everyone is noticing that you look different (don't all teens!). But here's a secret: Some teens are so worried about how they look and act they don't notice much about other people. It's part of being a teenager! Therefore, the best advice is to try to accept and be comfortable with yourself just the way you are. If you start to act like you are comfortable with yourself you may eventually really feel that way.

Ask your CF professional team if there are any adolescent support groups for chronically ill or chat rooms in your area. It helps to talk to other teens who are going through the same or similar things you are. They might have good ideas that you can use.

Embarrassment

There are some things about CF that may be embarrassing at times. Make sure your teachers and school personnel understand what they are – leaving the classroom as needed for bathroom visits, snacking in class and taking enzymes – and what they can do to make things easier. You and your parents should discuss any individual needs you may have with your teachers. A way to avoid some of this embarrassment is to educate your classmates about CF.

If you are comfortable enough to do this, you may do a class project about CF. This usually helps other teens understand what you are going through and why you may do some things differently. If you aren't comfortable sharing information with your entire class, you may start by sharing some information with close friends.

Bad Choices

Some of your friends may begin to make poor choices like drinking alcohol, smoking or using drugs. You might be tempted to try these things, too. You should know that doing these things have health consequences for you that they don't for your friends. **These consequences can be much more dangerous for you.** If your friends are pressuring you to smoke, drink alcohol or use drugs, you can choose to hang out with other friends who aren't messing around with such dangerous behaviors. If you are feeling pressured, you can use CF as a reason not to participate. These things can interact badly with your CF and the meds you take. They can be life-threatening consequences. If you do smoke, drink or use drugs and know you need to quit, ask your parents, Professional CF Self-Care team or a trusted adult for help. You are risking your life by participating in these activities. There are great resources to help you stop these behaviors.

Power Struggles

Common issues for teens with CF are parents who constantly ask questions and nag them to do their CF cares. What you don't realize is that your parents are worried about the consequences if you don't take good care of yourself. They are so used to taking care of you that it is hard for them to let go. If you haven't set up your self-care contract with your parents yet, now is the time to do it. This will show your parents that you want to take on more responsibility. It will help resolve daily battles over who does what and when.

Arguing with parents is typical for teens. However, if you feel like all you do is argue about your cares, your self-care contract should help eliminate much of this. If not, your CF team will have some good resources to help you find ways to compromise and problem-solve.

Strong Feelings

You may sometimes feel like you are on an emotional rollercoaster or that you have no control over your feelings. This is normal for teens. However, there are times when you should start to worry. If you begin to have intense feelings that don't change over time no matter what you are doing or that last more than a few weeks, they may not go away on their own. For instance, if you feel sad when you are sick, that is usually normal, but if you feel sad when you are hanging out with your friends, that may not be normal. It is a good idea to get used to checking in with yourself to see how you are feeling. If you notice you are having long periods of sadness, anger, hyperactivity, worry, anxiety, or you don't feel anything at all, you should seek help. It is important to use your support people to share your feelings. Whatever you do, tell someone. It won't help to keep these feelings inside. They won't go away on their own. Talking often helps you to get your feelings out. And getting your feelings out will make you feel better.

Doing too Little or too Much

Life is all about balance. If you do too little to keep yourself healthy you will probably get sick and if you do too much, and become obsessive about your CF cares, that is not healthy either. The key is to find the right balance that works for you. Communication is one of the best ways to find this balance. If you don't talk, no one will know how you are thinking and feeling. Look around at all the support people you have to help you through the rough times and to celebrate the good times. Use them. They agreed to be support people because they care about you and they want to be helpful and useful.

IV. The Future

A. Getting Ahead in the Real World

As you reach the latter part of your teenage years, you are able to think independently about your future. If the adults in your life, including your parents, members of your CF teams, teachers, counselors and friends, have done their jobs well, you have passed the stage of trying to convince everyone that you are an adult and are beginning to prepare yourself for your future. Let's look at some of the specific challenges of preparing for adulthood when you have CF.

To move into adulthood successfully, it is important to have a sense of optimism about the future, but it is even more important to have a feeling that one's life ahead truly will be worth living. The most difficult part of this challenge will be to allow for this sense of optimism within you and your family, no matter how mild or severe your CF symptoms happen to be. There have been wonderful advances in the care and treatment of individuals with CF in the past few decades and the research taking place is incredibly exciting. More and more individuals with CF will reach adulthood and go on to have satisfying lives filled with careers, families and friends and whatever else is on their dream list. How you go about this is key to being successful.

B. Achieving Your Goals

The goal is to have reached this bridge to adulthood understanding that your life can go one of two ways. There is the potential to develop severe disease (that could lead to a transplant or premature death) or the potential for good health (that could allow for a longer, fuller life). It is this understanding that will encourage you to hopefully reach for your dreams. Learning how to move ahead and work toward these dreams while also taking the time and energy needed for your CF self-cares is what most fulfilled adults with CF believe is the key to their success and happiness. This is a critical point. Reaching for your dreams, while balancing your self-cares, is the recipe for success!

One of the most important decisions of your life right now has to do with school and preparing for a career that will provide you with the income and benefits needed when one has CF. A job that includes, of course, health insurance but also sick and vacation leave (sometimes needed for tune-ups), short- and long-term disability, and a 401(k) or other retirement plan is obviously the best route. Many companies offer benefits for both full- and part-time employment. This means that college or vocational school is the route that will lead you to the best opportunities. Therefore, if you are still in high school, it will

be important for you to work toward getting grades good enough for college or vocational school acceptance. If you have dropped out of high school you should look at getting your GED. Or you can enroll through your community college in a curriculum that can lead to receiving your high-school diploma.

Sit down with your parents to discuss your goals for school and plan how to achieve them. Then feel free to explore these issues with your CF social worker and/or the people at your state's Division of Vocational Rehabilitation (DVR). They are often a valuable resource both for financial assistance and career counseling for people with CF and other medical conditions.

Tuition for any educational choice after high school can be very expensive and usually presents a difficult funding situation. Multiple scholarships are available, some exclusively for students with CF. Again, use your CF center social worker for assistance with scholarships and almost every other issue facing young adults with CF.

If attending college away from home is a goal, meet with your Professional CF self-care team to discuss if this is a realistic option for you. For many people with CF who have this desire, it truly is! However, there will be a number of challenges facing you. Life in a college dorm or apartment is a huge adjustment for most healthy college students since there are o parents around to set limits. For young adults with CF, it will take extra will power and determination to work CF cares into your schedule along with classes and college social life. In addition, potential absences for sick days and even courses of IV therapy may need to be addressed. Be sure you have informed any future roommates about your CF and what you need to do to care for yourself. For some, a single room with no roommates to worry about is the way to go. For all older adolescents and young adults with CF, the need to **balance your life** is a key point.

Many of you will be working to support yourself and pay for college tuition. This puts another stress on time that tempts one to miss ACT or other CF cares in order to go to class, sleep, work or socialize. You need to be disciplined about getting your CF cares done **FIRST** because if you don't, eventually you won't be well enough to go to class, work or socialize. Even though you think "it won't happen to me," it can. So be smart and place taking care of your CF at the top of your priority list. There are several CF center professionals including the CF social worker, nurse specialist and psychologist who can work with you to balance your life activities with your self-cares. This will be an ongoing issue that will need to be readjusted as time goes by and things in your life change. Keep in mind that anything is doable as long as you work to keep yourself well.

There is also the possibility for Social Security Disability when looking at options available to you. If full-time education or work seems like more than your health will allow you to handle, it's important for you and your parents to understand that qualifying for Disability does not mean that you are totally disabled. You can use your Disability Income to help you financially as you continue to think and plan for a life where part-time work or schooling is leading you to a satisfying life experience.

Social Security Disability can also provide you with the medical insurance that you may not be able to acquire through part-time employment. Remember, each person with CF has different capabilities and limitations and each person has the ability with the proper support to achieve a lot in all areas of life including education, employment, relationships, social activities, sports and much, much more.

C. Adult CF Care

The transition to an adult CF center is an important part of successful development during these late-teen and young-adult years. There has been considerable debate about whether patients and their families should switch from the pediatric CF team who has been following them for years, to a new team of adult medical providers. From a developmental and psychosocial perspective, there is no question that the move to an adult CF team makes the most sense.

It may sometimes be more comfortable for your parents, your pediatric team, and even you to maintain your healthcare with the team you've always known. However, having a team well versed in caring for the adult body with adult concerns makes the most sense since our goal is for adults with CF to live for decades. It is now the norm in most CF centers for the pediatric CF team to inform newly diagnosed families that in late adolescence their child's care will be transitioned to the adult CF team. This lets parents know right from the start that it is expected that their child will grow up and reach adulthood. This, then, should be their goal, too.

D. Adult Life

Young adults with and without CF are at the age when they start having serious relationships. More adults with CF are getting married and having families. These are issues to be thinking about, especially when making college and career decisions. Do you want to get married and have a family? If so, you will need a career that will support you and your future family.

Although the majority of men with CF are considered infertile, we now know that many do make sperm, but it is trapped in the testes (which are in the scrotum). For men with CF who would like to biologically have their own children, the technology now exists to retrieve their trapped sperm and then artificially inseminate it into their partner.

As mentioned earlier, women with CF are fertile and have been having healthy pregnancies for some time. It is a common misconception that they can't, and this is not true. Some women with CF have difficulty conceiving because of the thick mucus in the cervix and others do not. It is important that lung function and nutritional status are at acceptable levels before a woman with CF becomes pregnant. This will help facilitate a healthy pregnancy. A frequent question is whether ACT, especially the vest, will hurt the fetus – the answer is no.

Women with CF should talk to their healthcare provider about birth control and which method is right for them before becoming sexually active. They need to remember they can become pregnant, and this should be a well-planned, happy occasion – not an unexpected, unwanted surprise.

It is advisable that sexually active men and women practice safe sex (protecting yourself against undesired pregnancy and sexually transmitted diseases) by using condoms or other barrier methods. Before planning a pregnancy, all couples should have the non-CF partner tested for the CF gene. Remember, CF is a "recessive" genetic disease and requires a CF gene from both the mother and father for the child to have CF.

E. Resources for Real Life

As you become an adult, you will realize that there may be obstacles in many areas of life: education, employment, healthcare, housing, etc. However, there are many resources to help you, starting with your CF care center. There are many players on that team to help with specific areas, but the social worker is your key resource specialist. Not only can they help you problem-solve, but they can also provide resources that you may need to help make important life decisions. They are also available when you just need someone to talk to.

The CF Foundation is another powerful resource, which consists of both a national office and many state and local chapters. Check out their website at

http://www.cff.org for valuable information on education, research and fundraising activities. A major resource they helped to create for CF care centers is the *The Advocacy Manual: A Clinician's Guide to the Legal Rights of People with CF*. It is full of valuable information on laws affecting health insurance coverage, legal protection for students with CF, rights of employees with CF, and federal and state government benefit programs.

Another resource available at most colleges is the office for disability which can be helpful for a variety of issues from parking to class scheduling and housing concerns. Don't be afraid to check this out. It might be difficult to think you are "disabled." However, by simply having CF, you meet the legal definition of someone living with a disability. Using these resources is the best way to help you save energy, balance your life and reach your goals.

An important skill for all adults, but especially for those with chronic illness, is effective communication. If you have accomplished all that has been discussed in this booklet, you will already have achieved this. Setting up your CF self-care teams and self-care contract takes considerable skills in effective communication. You will need to be educating people about CF throughout your life – new friends, coworkers, bosses and significant others. This all depends on effective communication.

Having mature discussions with your CF care providers regarding healthcare decisions also involves effective communication. This skill is an invaluable tool for all aspects of your life and your future. If you feel your communication skills are less than desirable, talk to your CF social worker about resources that can help you develop better communication skills.

F. Leaving the Best for Last

Thankfully, we have now reached a time where living into one's 30s with CF is nothing exceptional and, given advances in medical care, living into your 50s and 60s, for many, is now a reasonable goal. Therefore, everyone – parents, teens and team members – must understand that creating the strength and independence required for this potentially long journey through life with CF must become an important priority, beginning at the moment of diagnosis.

We must all make a collaborative effort to achieve this. We must also work together to push our state and local governments to provide for the medical care, education and legal rights that all people with CF need and deserve. Hopefully, you can learn to make good decisions as you seek healthcare, relationships, careers and other aspects of a long and fulfilling life, which you deserve.

We hope this booklet is an important resource for helping you succeed as you move from your teenage years to your young adult years. Each of us is only given one body to take us through this journey called life. Your health is your most important and most precious commodity. The most effective way to help you protect and improve this precious commodity, your health, is to learn self-care.

We wish you the best of everything and hope you reach for your dreams.

V. Resource Section

Table I: CF Self-Care Questionnaire

Developed by Mary Jo McCracken, RN, MS, CPNP, Pediatric Pulmonary Nurse Practitioner and Clinical Nurse Specialist

Name_____ Date_____ Age_____

Directions: The purpose of this test is to measure how much you know about Cystic Fibrosis (CF), its complications and treatments. Read each question carefully. Answer each question as completely as you can. Each question is worth a different number of points. The number of points you can receive for each question depends upon how many correct details or how completely you can answer each question. Therefore, write down as many correct details or as much as you know that can answer that question. The total amount of points possible for each question is listed.

1) How does an individual get CF? (3 possible points)

2) In general, describe how CF affects your body OVERALL (not specific body parts). (6 possible points)

3) In picture at the end of this questionnaire, draw in the lungs, heart, pancreas, stomach and small intestines where they belong. Label each body part drawn. (5 possible points)

4) What do your lungs do for your body? (2 possible points)

5) What does your pancreas do for your body? (3 possible points)

6) What happens to food in your intestine? (3 possible points)

7) How can CF cause problems in your lungs? (10 possible points)

8) How can CF cause problems in your pancreas? (6 possible points)

9) What are the problems CF can cause in your intestine? (6 possible points)

10) What problems could happen if your body does NOT have enough digestive enzymes? (8 possible points)

11) How can CF cause problems with your sweat? (2 possible points)

12) What treatment(s) can your CF healthcare provider (CF HCP) prescribe for the CF problems in your lungs? (8 possible points)

13) What treatment(s) can your CF HCP prescribe for the CF problems in your pancreas? (4 possible points)

14) What treatment(s) can your CF HCP prescribe for the CF problems in your intestines? (4 possible points)

15) Name 3 medications you are taking and explain how they help your body. (There are 6 possible points for this question, 1 for each medication named and 1 for how each helps your body.)

16) What kinds of food should a person with CF eat a lot of? (3 possible points)

17) Explain why nutrition is important for an individual with CF. (5 possible points)

18) Tell how each of the items below can be helpful in the treatment of CF.

a. Aerosol treatments. (5 possible points)

b. Airway Clearance Therapies (chest physical therapy, CPT, BD, the Vest, flutter, PEP device, autogenic drainage). (5 possible points)

c. Coughing/huffing. (3 possible points)

d. Antibiotics. (1 possible point)

19) Give 3 reasons why exercise is important for a person who has CF. (3 possible points)

20) How often should your aerosol equipment be cleaned? (2 possible points)

21) Explain how you should clean and disinfect your aerosol equipment. (7 possible points)

22) Why is it important to clean your aerosol equipment? (2 possible points)

23) What is the correct solution for your aerosol treatment? (4 possible points)

24) What can you do to prevent becoming low in body salts? (3 possible points)

25) Name 3 symptoms of heat exhaustion. (3 possible points)

26) Name 2 possible complications of CF in the upper respiratory system (above the lungs; ears nose sinuses throat). (2 possible points)

27) For each complication you listed in question #26, describe the appropriate treatment. State the complication and its treatment. (2 possible points)

28) Describe how the lungs naturally clean themselves. (4 possible points)

29) Describe the steps that should take place with your ACT's (airway clearance therapies). Choose only the therapy you use and describe the steps involved.
P&PD or CPT (5 possible points), Vest (11 possible points), Positive Expiratory Pressure (PEP) (5 possible points), Active Cycle of Breathing (ACBT) (6 possible points), Autogenic Drainage (AD) (4 possible points), acapella® (9 possible points), Flutter (7 possible points)

30) How are straight digestive enzyme supplements different from enteric coated digestive enzyme supplements? (3 possible points)

31) How would you know if you were not taking enough digestive enzymes with your food? (8 possible points)

32) Describe the proper way to breathe during an aerosol treatment. (3 possible points)

YAY! YOU ARE DONE! GREAT JOB!

Table II: CF Self-Care Questionnaire Answer Key

Developed by Mary Jo McCracken, RN, MS, CPNP, Pediatric Pulmonary Nurse Practitioner and Clinical Nurse Specialist

Answers included after question. Total Points: 146-153

Total points depend on your answer for question #29. In this question, there are different numbers of points for each form of ACT.

Name_____ Date_____ Age_____

Max Points Available

(3 pts) **1)** How does an individual get CF?
a) a gene or inherited (1 point)
b) from both parents (1 point)
c) recessive gene (1 point)

(6 pts) **2)** In general, describe how CF affects your body OVERALL (not specific body parts).
a) problem/mutation with CFTR (CF transmembrane regulator) (1 point)
b) problem/defect with ion transport/problem with sodium and chloride in and out of exocrine cells or exocrine cells do not function properly (1 point)
c) decreased airway surface fluid (1 point)
d) mucus is thick, sticky or dehydrated (1 point)
e) body parts get obstructed/clogged up/plugged up (1 point)
f) sweat has more salt than normal (1 point)

(5pts) **3)** In the picture at the end of this Questionnaire, draw in the lungs, heart, pancreas, stomach and small intestines where they belong. Label each body part drawn. See picture at end of Answer Key for the correct drawing.

(2 pts) **4)** What do your lungs do for your body?
a) exchange gases (takes in O and releases CO) (2 point)
b) breathing (1 point)
c) takes in oxygen (O) (1 point)
d) removes carbon dioxide (CO) (1 point)
*maximum of 2 points for this question – 2 points for “a” and 1 point for “b”, “c”, and “d”.

(3 pts) **5)** What does your pancreas do for your body?
a) makes digestive enzymes/enzymes/helps digest food (1 point)
b) makes insulin (1 point)
c) give extra point for listing specific digestive enzymes-lipase, amylase, protease (1 point)

(3 pts) **6)** What happens to food in your intestine?
a) is digested (1 point)
b) is absorbed (1 point)
c) waste products are stored and eliminated (1 point)

(10 pts) **7)** How can CF cause problems in your lungs?
a) mucus is thick/stickier (1 point)
b) passages/airways/bronchi get obstructed/plugged up (1 point)
c) increases work of breathing/harder to breathe (1 point)
d) can not get proper O /CO exchange (1 point)
e) bacteria grow/develop infection (1 point)
f) have swelling/inflammation in airways/bronchi (1 point)
g) can scar and destroy lung tissue (1 point)
h) can develop spasms in airways/passages/bronchi (bronchospasm) (1 point)
i) develop bronchiectasis (1 point)
j) alveoli collapse/blebs form and can rupture/pneumothorax (1 point)

(6 pts) **8)** How can CF cause problems in your pancreas?
a) passages/ducts get obstructed/plugged/clogged (1 point)
b) thick mucus clogs/obstructs/plugs passages/ducts (1 point)
c) digestive enzymes can’t get out/insufficient enzymes (1 point)
d) food doesn’t get digested properly (1 point)
e) diabetes/problems with insulin (1 point)
f) pancreatitis (1 point)

(6 pts) **9)** What are the problems CF can cause in your intestine?
a) thick mucus causes problems (1 point)
b) can have decreased food digestion and absorption (1 point)
c) can't absorb fat soluble vitamins properly (1 point)
d) can get a blockage/obstruction/mucus ball/distal ileus obstructive syndrome or DIOS (1 point)
e) rectal prolapse (1 point)
f) intussusception/telescoping of intestine (1 point)

(8 pts) **10)** What problems could happen if your body does NOT have enough digestive enzymes?
a) undigested food (1 point)
b) poor absorption of food (1 point)
c) poor growth (1 point)
d) more frequent bowel movements/stools (1 point)
e) stools float/greasy stools (1 point)
f) cramps/gas/distended abdomen/bloating (1 point)
g) rectal prolapse (1 point)
h) DIOS (distal ileus obstructive syndrome) (1 point)

(2 pts) **11)** How can CF cause problems with your sweat?
a) has too much salt (1 point)
b) can become low in body salts (1 point)

(8 pts) **12)** What treatment(s) can your CF healthcare provider (CF HCP) prescribe for the CF problems in your lungs?
a) aerosol treatments/saline and bronchodilator (1 point)
b) mucolytics such as Pulmozyme®/Mucomyst® (1 point)
c) airway clearance therapies/chest physical therapy/the Vest/autogenic drainage/Flutter/Active Cycle of Breathing/PEP (1 point)
d) exercise (1 point)
e) antibiotics; oral, IV, aerosols (1 point)
f) breathing exercises/cough/huff (1 point)
g) bronchodilators; oral, MDI or aerosol (1 point)
h) other medications; steroids, expectorants, anti-inflammatory meds, etc. (1point)

(4 pts) **13)** What treatment(s) can your CF HCP prescribe for the CF problems in your pancreas?
a) digestive enzyme supplements/enzymes (1 point)
b) insulin (1 point)
c) bile salts (1 point)
d) antacids/cimitadine/zantac (1 point)

(4 pts) **14)** What treatment(s) can your CF HCP prescribe for the CF problems in your intestines?
a) digestive enzyme supplements/enzymes (1 point)
b) fat soluble vitamins (1 point)
c) high calorie, high protein diet (1 point)
d) drink Mucomyst®/Miralax®/Golytely® (1 point)

(6 pts) **15)** Name 3 medications you are taking and explain how they help your body.
- give 1 point for each medication listed and 1 point for each correct mode of action for medication listed.

(3 pts) **16)** What kinds of food should a person with CF eat a lot of?
a) high calorie/high fat food (1 point)
b) high protein (1 point)
c) salty foods (1 point)

(6 pts) **17)** Explain why nutrition is important for an individual with CF.
a) to grow properly/gain weight (1 point)
b) to help resist/recover from illness (1 point)
c) you use/need more calories (1 point)
d) digestion and absorption are a problem (2 points)
e) work of breathing/infection requires more calories (1 point)

18) Tell how each of the items below can be helpful in the treatment of CF.
(5 pts) **a)** Aerosol treatments:
1) thins/loosens mucus/breaks up mucus (1 point)
2) decreases spasms in airways/dilates airways (1 point)
3) give additional point if specific medication is named (1 point)
4) antibiotic aerosols treat bacteria/infection in lungs (ie: Tobi®) (1 point)
6) Pulmozyme® breaks up mucus, makes mucus easier to cough up (1 point)

(5 pts) **b)** Airway Clearance Therapies (chest physical therapy, bronchial drainage, the Vest, the Flutter, positive expiratory pressure device (PEP), autogenic drainage, Active Cycle of Breathing, acapella®):
shakes mucus loose (1 point)
helps move/cough up mucus (1 point)
improves breathing, gas exchange, O , CO in lung (1 point)
decreases amount of mucus in lung/clogging airways (1 point)
removes infected pus, bacteria/fights infection (1 point)

(3 pts) **c)** Coughing/huffing:
1) moves/removes mucus from lung (1 point)
2) improves breathing/gas exchange (1 point)
3) fights infection/removes infected pus, bacteria (1 point)

(1 pt) **d)** Antibiotics:
kills germs/bacteria, fights infection/bacteria, prevents infection (1 point)

(3 pts) **19)** Give 3 reasons why exercise is important for a person who has CF.
a) helps move mucus up and out of lungs (1 point)
b) strengthens muscles of breathing (1 point)
c) improves vital capacity/lung function (1 point)

(2 pts) **20)** How often should your aerosol equipment be cleaned?
a) after every use (1 point)
b) once a day (1 point)

(7 pts) **21)** Explain how you should clean and disinfect your AEROSOL equipment.
a) wash your hands (1 point)
b) the neb parts must be cleaned before they can be disinfected. Therefore, with a clean paper towel, wash the inside and outside of neb parts with liquid soap and hot water. Rinse well. (1 point)
c) then, disinfect neb parts using one of the following methods: (give 1 point for each correct method given)
1. boil for 5 minutes on stove, cool completely. Beware of melting neb parts. (1 point)
2. dishwasher – place in upper rack. Water temperature must be at least 158 degrees for 30 minutes. Beware of melting parts. (1 point)
3. Microwave in water for 5 minutes. (1 point)
4. The CF Foundation also includes: soak in one of the following solutions. (1 point – must have correct solution to earn point)
-1 part bleach to 50 parts water for 3 minutes
- 70% isopropyl alcohol for 5 minutes
- 3% hydrogen peroxide for 30 minutes.

**Rinse well with sterile water.
These solutions can be very caustic to the mouth and lung if not thoroughly rinsed after soaking. Therefore, it is our recommendation NOT to use these solutions. However, discuss cleaning options with your CF HCP and Respiratory Therapist for their recommendations.
d) place disinfected neb parts on clean paper towel and cover with another clean paper towel until next use. (1 point)

(2 pts) **22)** Why is it important to clean your aerosol equipment?
a) to prevent medication build-up (1 point)
b) to prevent mold/bacteria growth (1 point)

(3 pts) **23)** What is the correct solution for your aerosol treatment?
a) give 1 point for each medication named correctly (2 points)
b) give 1 point if amount of medication is given (1 point)

(3 pts) **24)** What can you do to prevent becoming low in body salts?
a) drink electrolyte drinks (ie: Gatorade®) (1 point)
b) eat salty foods (1 point)
c) add salt to food (1 point)

(3 pts) **25)** Name 3 symptoms of heat exhaustion.
a) weakness (1 point)
b) dizziness (1 point)
c) headache (1 point)
d) muscle cramp (1 point)
e) increased sweating (1 point)
f) cool, clammy skin (1 point)
g) nausea/vomiting (1 point)
h) lethargy/exhaustion/fatigue (1 point)
i) confusion (1 point)
j) convulsions (1 point)

(2 pts) **26)** Name 2 possible complications of CF in the UPPER respiratory system (above the lungs) (ears, nose, sinuses, throat)
a) nasal polyps (1 point)
b) ear infection (1 point)
c) clouding of sinuses/mucus in sinuses (1 point)
d) sinusitis/infection of sinuses (1 point)

(2 pts) **27)** For each complication listed in the above question, describe the appropriate treatment. State complication and its treatment.
a) nasal polyps – nasal spray (1 point)
b) nasal polyps – surgical removal (1 point)
c) ear infection – antibiotics (1 point)
d) sinusitis – nasal sprays (1 point)
e) sinusitis – antibiotics (1 point)

(4 pts) **28)** Describe how the lungs naturally clean themselves.
a) goblet cells make mucus/mucus lines the airways/bronchi (1 point)
b) mucus traps dirt, bacteria, etc... (1 point)
c) cilia moves mucus upward (1 point)
d) mucus is coughed out or swallowed (1 point)

(4-11 pts) **29)** Describe the steps that should take place with your ACT (airway clearance therapies).
Choose only which ACT you use and score your answer
a) using P and PD or CPT (5 possible points)
1) aerosol before clapping (1 point)
2) clapping/pounding (1 point)
3) vibrating(1 point)
4) cough/huff between positions (1 point)
5) change positions and repeat 1-4 (1 point)
b) Using the VEST (11 possible points)
1) aerosol treatments done during Vest (1 point)
2) put Vest jacket on, set machine(compressor) to a pressure of 10 and begin first frequency which is usually 8 (1 point for pressure and 1 point for frequency)
3) each frequency should be done for 5 minutes (1 point)
4) after holding frequency for 5 minutes deflate vest and huff/cough 3 times (1 points)
5) move frequency to 9 at same pressure and hold for 5 minutes, then deflate and huff/cough 3 times (1 point)
6) move frequency to 10 and hold for 5 minutes then deflate and huff/cough 3 times (1 point)
7) change pressure on compressor to 6 and change frequency to 18 and hold for 5 minutes then deflate and huff/cough 3 times (1 point for pressure and 1 point for frequency)
8) move frequency to 19 and hold for 5 minutes then deflate and huff/cough 3 times (1 point)
9) move frequency to 20 and hold for 5 minutes then deflate and huff/cough 3 times (1 point)
c) Positive Expiratory Pressure Therapy (PEP) (5 possible points)
1) if using with aerosol, set that up and check PEP device or resistor for proper setting (1 point)
2) breathe in normally, exhale against resistance from PEP device (1 point)
3) every 10-20 breaths you should huff/cough (1 point)
4) huff/cough several times and repeat 2 and 3 (1 point)
5) should continue for 20-30 minutes (1 point)
d) Active Cycle of Breathing (ACBT) (6 possible points)
Combination of cycles of:
1) breathing control – normal, gentle breathing with the lower chest (1 point)
2) thoracic expansion exercises – deep breaths with breath holding (1 point)
3) forced expiration technique (huff/cough) - huffs with breathing control (1 point)
*Give an additional point for each cycle that is correctly named.
e) Autogenic Drainage (AD) (4 possible points)
consists of a cycle of breathing through 3 lung volumes which aid:
1) unsticking /dislodging mucus – low lung volume breathing (1 point)
2) collecting mucus – breathing at slightly larger volumes (1 point)
3) evacuating/clearing mucus – normal to high lung volume breathing (1 point)
4) huff/coughs (1 point)
f) acapella® (oscillating PEP) (9 possible points)
1) if using with aerosol set that up and attach to acapella®(1 point)
2) slowly take a breath in that is slightly bigger than normal and hold your breath for 2-3 seconds (1 point)
3) place acapella® mouthpiece in your mouth or if using a mask, hold it over your mouth and nose creating a seal and exhale fast but not forcefully through the acapella® (1 point)
4) tune the acapella® to achieve the best vibration (1 point)
5) repeat 2 and 3 five to ten times (1 point)
6) after completing 5-10 breaths (steps 2-5) take a deep breath in (as deep as you can) and hold it for 2-3 seconds (1 point)

7) put the acapella® mouth piece in your mouth (or mask over mouth and nose) and exhale forcefully until all the air is out of your lungs (1 point)
8) huff/cough – repeat 2-3 times (1 point)
9) repeat all above steps for 15-20 minutes (1 point)

g) The Flutter (7 possible points)
1) slowly take a breath in that is slightly bigger than normal and hold your breath for 2-3 seconds (1 point)
2) place flutter mouth piece in mouth and exhale fast but not forcefully through the flutter (1 point)
3) repeat steps 1and 2 five to ten times trying not to cough (1 point)
4) after completing step 3, take a deep breath in (as deep as you can) and hold it for 2-3 seconds (1 point)
5) put the flutter mouthpiece in your mouth and exhale forcefully until all the air is out of your lungs (1 point)
6) huff/cough – repeat 2-3 times (1 point)
7) repeat all above steps for 15-20 minutes (1 point)

(3 pts) **30)** How are straight digestive enzyme supplements different from enteric-coated digestive enzyme supplements? (3 possible points)
a) straight enzymes – start working right away/are not time release/are not coated (1 point)
b) enteric-coated enzymes – are time released beads/start to work in small intestine/bowel (1 point)
c) enteric-coated enzymes are stronger than straight enzymes /digest food better (1 point)

(8 pts) **31)** How would you know if you were not taking enough digestive enzymes with your food? (8 possible points)
a) have more than 2 bowel movements/stools a day (1 point)
b) stools would float/greasy stools (1 point)
c) abdominal cramps (1 point)
d) excessive gas (1 point)
e) poor weight gain/poor growth (1 point)
f) rectal prolapse (1 point)
g) DIOS (distal intestinal obstruction syndrome) (1 point)
h) Stools loose, undigested food in them (1 point)

(3 pts) **32)** Describe the proper way to breathe during an aerosol treatment. (3 possible points)
a) slow breathing (1 point)
b) deep breathing (1 point)
c) breath holding (1 point)

CONGRATULATIONS! YOU ARE DONE!

NOW SCORE YOURSELF:

PhD level .....138-153 points

Masters Degree .....120-137 points

College Degree .....100-119 points

High School Degree .....70-99 points

Less than 70 points .....You need to spend some quality time learning more about CF. Ask your CF Professional Self-Care Team for help.

Table III: The CF Body Check Log

Use the rating scale below to answer the following questions:

Each section will ask you to rate the status of that body part, as well as, answer different questions. *For each of these – circle the correct number or response.*

1	2	3	4	5	6	7	8	9	10
excellent	very good		good/average		fair		poor	sick	worst ever

Date: _____

Overall:	- Status? 1 2 3 4 5 6 7 8 9 10 - Are you better/worse/same as yesterday? - If worse, why?
Check appearance:	- Face? (swelling, sunken eyes, rash, bruising). - Color? (pink, pale, dusty, blue-tinged lips). - Fever? (yes/no)
Sleep:	- Status? 1 2 3 4 5 6 7 8 9 10 - How does this compare to the night before? (better/worse/the same) - Did cough/shortness of breath/post-nasal drip interrupt your sleep? (yes/no) - Are you able to sleep lying flat? (yes/no) - If there is a change in your sleep what do you need to do about it?

Upper Respiratory System

Nose:	- Problem? (yes/no) - Status? 1 2 3 4 5 6 7 8 9 10 - What was the status yesterday? - If worse, what should you do about it? - Can you breathe through your nose? (yes/no) - Nasal secretion? (yes/no) - If yes, what color (❑ clear/white, ❑ yellow, ❑ green, ❑ bloody)? - Is this a change? (yes/no) - Amount of secretions? __tsp(s), __TBSP(s), __cup(s) - Is this more/less/no change?
Sinus:	- Is there a problem? (yes/no) - Status? 1 2 3 4 5 6 7 8 9 10 - Is this better/worse/same as yesterday? - If worse, what should you do? - Do you have a headache? (yes/no) - If yes, what is the status? 1 2 3 4 5 6 7 8 9 10 - Do you need to do anything about your headache? **Headaches, especially when first getting up in the a.m., need to be monitored.**

Mouth:	- Color of lips (pink, pale, bluish/dusky). - Is this a change? (yes/no) - Do you have mouth sores or problem teeth? (yes/no) - How is your throat (red, swollen, sore)? - Do you have foul breath or a foul taste in your mouth? - Status of mouth? 1 2 3 4 5 6 7 8 9 10 - Is this a change from yesterday? (yes/no) - If yes, what should you do? **Foul breath should be monitored!**

Lower Respiratory System

Lungs:	- Respiratory Rate? #_____min. - Is this a change from your baseline? (yes/no)
Cough:	- Has your cough changed? (yes/no) If yes, how? - How often do you cough? - Amount: ❑ none, ❑ rarely, ❑ some days, ❑ most days, ❑ every day, ❑ only with CPT or exercise. - Frequency: ❑ none, ❑ 1-2x/12 hours, ❑ 1-2x/6 hours, ❑ 1-2x/2 hours, ❑ 1-2x/hour, ❑ many times every hour. - Is this cough productive (❑ no sputum, ❑ clears throat/swallows, ❑ sometimes productive, ❑ always productive)? - What makes you cough (❑ ACT, ❑ laughing, ❑ exercise, ❑ crying, ❑ colds, ❑ tickling, ❑ aerosols, ❑ smoking)? - Using the following scale, what is your status? 1 (none), 2 (clears throat), 3-4 (mild), 5-6 (moderate), 7-8 (a lot), 9 (severe), 10 (worst ever). - Circle your correct number: 1 2 3 4 5 6 7 8 9 10 - Is this a change? (yes/no) - If yes, what should you do about it? **BEWARE OF SUPPRESSING YOUR COUGH!**
Sputum:	- Amount of sputum (none, clears throat and swallow, ____tsp(s), _____TBSP(s), ____cup(s). - Color of sputum (❑ clear/white, ❑ yellow, ❑ green, ❑ brown, ❑ bloody). - Consistency of sputum (❑ thin, ❑ thick, ❑ thicker, ❑ very thick). - Using the following scale, what is your status? 1 2 3 4 5 6 7 8 9 10 - Has your sputum changed? (yes/no) If so, what should you do about it?
Shortness of Breath (SOB):	- Do you experience shortness of breath (yes/no)? - Using the following scale, what is your status? 1 2 3 4 5 6 7 8 9 10 - Is this a change from yesterday (yes/no)? - If yes, is it better or worse? - If worse, what should you do about it?
Wheeze:	- Are you experiencing any wheezing? (yes/no) - If yes, when do you wheeze (❑ with light exercise, ❑ with heavy exercise, ❑ at rest, ❑ cold air, ❑ smoke, ❑ coughing, ❑ other)? - Using the following scale, what is your status? 1 2 3 4 5 6 7 8 9 10 - How often do you wheeze (❑ rarely, ❑ some days, ❑ most days, ❑ everyday, ❑ several times per day, ❑ once every few hours, ❑ every hour, ❑ several times an hour)? - Is this better/worse/the same as yesterday? - If worse, what should you do about it?

Exercise:	- What is your ability to be physically active (❑ unlimited, ❑ average, ❑ rests a bit, ❑ rests a lot)? - Using the following scale, what is your status? 1 2 3 4 5 6 7 8 9 10 - Is this a change? (yes/no) - If yes, is it better or worse? - If worse, what should you do about it?
Chest Pain:	- Do you have any chest pain? (yes/no) - If yes, where is it? - Is there pain with inspiration, expiration or both? - Using the following scale, how severe is the pain? 1 2 3 4 5 6 7 8 9 10 - Is this new pain? (yes/no) - Has the pain gotten worse/better/stayed the same? - What makes the pain better or worse? - Do you have limited chest movement or do both sides move equally? - Do you need to do anything about any chest pain?
Gastrointestinal System	- Weight today: _____lbs -Weight goal: _____lbs - Weight at last weigh-in: _____lbs - Are you meeting your weight goal (yes/no)? - If no, why not? Write down your answers
Appetite:	- Using the following scale, how is your appetite? 1 2 3 4 5 6 7 8 9 10 - Is this a change/ (yes/no) - Is it better/worse/the same? - If worse, why? - Any nausea or vomiting? (yes/no) - If yes, how much and how often? - What makes nausea and/or vomiting better or worse? - Do you have any heartburn or reflux? (yes/no) - If yes, what is the status? 1 2 3 4 5 6 7 8 9 10 - Is this better/worse/the same as yesterday? - If worse, what should you do about it?
Abdomen:	- Do you have any abdominal pain? (yes/no) - If yes, where is it and how severe is it? 1 2 3 4 5 6 7 8 9 10 - Is this pain new? (yes/no) - How long have you had this pain? - Is the pain better, worse or the same as yesterday? - What makes the pain better or worse? - Are you pooping regularly (bowel movement, stool)? (yes/no) - If no, when was your last BM/stool? - Do you have any abdominal bloating? (yes/no) - Gassiness? (yes/no) - Do you need to do anything about your abdominal complaints?
Stools:	- How many stools did you have yesterday (1, 2, 3, 4, more)? - Are they usually: ❑ normal, ❑ greasy, ❑ fatty, ❑ gassy, ❑ floating, ❑ snake-like, ❑ diarrhea, ❑ constipation, ❑ rectal prolapse. - If your stools are not what they should be, what do you need to do? - Stools should sink, not float. **Watch for signs of DIOS.**

Extremities:	- Any swelling of ankles, feet, fingers or hands? (yes/no) - If yes, where is the swelling and how long have you had it? - Is the swelling better/worse/the same? - Do you bruise easily? - If yes, are you taking vitamin K regularly? - Using the following scale, what is your status? 1 2 3 4 5 6 7 8 9 10 - Do you need to do anything about it?
Pain:	- Any pain in your head, chest, stomach, abdomen, joints, other? - Is this new pain? (yes/no) - Status (rate your pain)? 1 2 3 4 5 6 7 8 9 10 - Is the pain better/worse/the same as yesterday? - What makes the pain better or worse? - Do you need to do anything about your pain?
Outlook:	- How is your mood? - Are you ❑ cheerful, ❑ content, ❑ happy, ❑ serious, ❑ variable, ❑ sad, ❑ tearful, ❑ irritable, ❑ angry, ❑ whiny, ❑ frustrated or ❑ depressed? - Is this a change? (yes/no) - Is your mood better or worse? - If worse, what do you need to do about it?
Other:	- This is for you to personalize if needed: glucose check, line care, g-tube care, etc. _____ _____ _____ _____ _____ _____ _____ _____ _____ _____ _____
End of Body Check:	- Overall status? 1 2 3 4 5 6 7 8 9 10 - Compare to beginning overall status, which was _____. - If they are significantly different, review your body check to figure out why.

YOU ARE DONE!
GREAT JOB!

Table IV: CF Self-Care Schedule

	Monday	Tuesday	Wednesday	Thursday	Friday	Saturday	Sunday
5 a.m.							
6 a.m.							
7 a.m.							
8 a.m.							
9 a.m.							
10 a.m.							
11 a.m.							
12 Noon							
1 p.m.							
2 p.m.							
3 p.m.							
4 p.m.							
5 p.m.							
6 p.m.							
7 p.m.							
8 p.m.							
9 p.m.							
10 p.m.							
11 p.m.							
12 p.m.							

Table V: CF Self-Care Contract

I, _____ (name), in an effort to become more responsible for my health, willingly agree to this self-care contract.

I will be responsible for: (list all CF cares you will do)	**Parents will be responsible for: (list all CF cares mom/dad will do)**
1. _____	1. _____
2. _____	2. _____
3. _____	3. _____
4. _____	4. _____
5. _____	5. _____

Reinforcers/Rewards:	**What is needed to earn the rewards:**
1. _____	1. _____
2. _____	2. _____
3. _____	3. _____
4. _____	4. _____
5. _____	5. _____

Signed

Teen_____ Date:_____

Parent(s)_____ Date:_____

Internet Resources: Informational Web Sites on Cystic Fibrosis

Here is a sample of informative websites on Cystic Fibrosis (CF). This list is not meant to be inclusive. It is simply a list of sites we have found helpful. Please keep in mind that you should NEVER change your CF treatment plan without first discussing it with your CF healthcare provider. Discuss information and/or new ideas you have found on the internet with your CF healthcare provider or team members. If there is a website on CF you think is especially helpful, please let us know. Send it to MJ McCracken at digestivecare@fast.net.

Use chat rooms with caution. Be careful with information you give out as well as information you receive.

CF Organizations

Boomer Esiason Foundation
www.esiason.org
A resource for those affected by CF. Includes details about this foundation, fund raising, events and a newsletter.

Cystic Fibrosis Foundation
www.cff.org
A wealth of information on CF and CF research. The standards of care are listed on this website.

Cystic Fibrosis Worldwide
www.cfww.org
Newsletter, links, updates on CF international platform for the exchange of information

International CF Adult Organization
www.iacfa.org
Organization for adults with CF throughout the world.
Very informative.

Cystic Fibrosis Medicine
www.cysticfibrosismedicine.com

Medical info on CF
www.respiratorycare.medscape.com
News and information on pulmonary issues.

My Cystic Fibrosis
www.mycysticfibrosis.com
Helpful information on CF.

Cystic Fibrosis Research Inc.
www.cfri.org
Help to fund and sponsor life saving CF research, provide educational programs and personal support.

CF Support Groups

Reach for the Stars
www.r4stars.org
Provides resources, knowledge and support to those with CF.

Cystic-L
www.cystic-L.org
Information and support. Free email service dedicated to the exchange of information and support on CF.

Pacer
www.pacer.org/
MN foundation for chronically ill patients and families

A CF community website
www.cysticfibrosis.com
"Support. Education. Hope." For those with CF.

Starbright Foundation
www.starbright.org
Provides education, family activities and entertainment to seriously ill children.

Breathing Room
www.theBreathingRoom.org
"The Art of Living with CF."

CF parents group
www.Groups.Yahoo.com/group/cfparents
"A place for parents to children with CF to come for support and information."

CF Scholarship Info

Scholarship Info
www.CFF.org/search_cff_org/search_results.cfm
Information on scholarships available.

National

Federal Government Program
www.usa.org
All information related to federal laws regarding insurance and education plans for children with special needs.

National Library of Medicine
www.nlm.nih.gov
Articles on CF research and national medical news.

Search Engine Sites

Google
www.google.com

Yahoo
www.yahoo.com

Notes

Footnotes

Golytely® is a registered trademark of Braintree Laboratories

MIC-KEY® is a registered trademark of Kimberly-Clark Worldwide, Inc.

Miralax® is a registered trademark of Schering-Plough Corp

Motrin® is a registered trademark of Johnson & Johnson Company

Mucomyst® is a registered trademark of Bristol-Myers Squibb

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