

Welcome To My Home

"Some see a hopeless end while others see endless hope" -Author Unknown



My Name Is Shira
And I have Spinal Muscular
Atrophy Type 1

SHIRA'S PROTOCOL DISCLAIMER

Please note that this protocol was developed specifically for Shira's care and that you should consult your health care provider before implementing any protocol to your child. While this plan works great for Shira it should not be depended on for other children. Each child must have there individual plan that caters to their direct needs. This plan includes some universal basic information but also contains some Shira specific strategies that could potentially be harmful to other children that require other specific procedures.

You should form your own plan by using the NIV Protocol of Dr. Bach, Dr. Schroth, RT Brian Weaver and Dr. Swoboda as well as your personal health care provider.

Hippocratic Oath -- Classical Version

I swear by Apollo Physician and Asclepius and Hygieia and Panacea and all the gods and goddesses, making them my witnesses, that I will fulfil according to my ability and judgment this oath and this covenant:

To hold him who has taught me this art as equal to my parents and to live my life in partnership with him, and if he is in need of money to give him a share of mine, and to regard his offspring as equal to my brothers in male lineage and to teach them this art - if they desire to learn it - without fee and covenant; to give a share of precepts and oral instruction and all the other learning to my sons and to the sons of him who has instructed me and to pupils who have signed the covenant and have taken an oath according to the medical law, but no one else.

I will apply dietetic measures for the benefit of the sick according to my ability and judgment; I will keep them from harm and injustice.

I will neither give a deadly drug to anybody who asked for it, nor will I make a suggestion to this effect. Similarly I will not give to a woman an abortive remedy. In purity and holiness I will guard my life and my art.

I will not use the knife, not even on sufferers from stone, but will withdraw in favor of such men as are engaged in this work.

Whatever houses I may visit, I will come for the benefit of the sick, remaining free of all intentional injustice, of all mischief and in particular of sexual relations with both female and male persons, be they free or slaves.

What I may see or hear in the course of the treatment or even outside of the treatment in regard to the life of men, which on no account one must spread abroad, I will keep to myself, holding such things shameful to be spoken about.

If I fulfill this oath and do not violate it, may it be granted to me to enjoy life and art, being honored with fame among all men for all time to come; if I transgress it and swear falsely, may the opposite of all this be my lot.

Translation from the Greek by Ludwig Edelstein. From The Hippocratic Oath: Text, Translation, and Interpretation, by Ludwig Edelstein. Baltimore: Johns Hopkins Press, 1943.

Hippocratic Oath—Modern Version

I swear to fulfill, to the best of my ability and judgment, this covenant:

I will respect the hard-won scientific gains of those physicians in whose steps I walk, and gladly share such knowledge as is mine with those who are to follow.

I will apply, for the benefit of the sick, all measures [that] are required, avoiding those twin traps of overtreatment and therapeutic nihilism.

I will remember that there is art to medicine as well as science, and that warmth, sympathy, and understanding may outweigh the surgeon's knife or the chemist's drug.

I will not be ashamed to say "I know not," nor will I fail to call in my colleagues when the skills of another are needed for a patient's recovery.

I will respect the privacy of my patients, for their problems are not disclosed to me that the world may know. Most especially must I tread with care in matters of life and death. If it is given me to save a life, all thanks. But it may also be within my power to take a life; this awesome responsibility must be faced with great humbleness and awareness of my own frailty. Above all, I must not play at God.

I will remember that I do not treat a fever chart, a cancerous growth, but a sick human being, whose illness may affect the person's family and economic stability. My responsibility includes these related problems, if I am to care adequately for the sick.

I will prevent disease whenever I can, for prevention is preferable to cure.

I will remember that I remain a member of society, with special obligations to all my fellow human beings, those sound of mind and body as well as the infirm.

If I do not violate this oath, may I enjoy life and art, respected while I live and remembered with affection thereafter. May I always act so as to preserve the finest traditions of my calling and may I long experience the joy of healing those who seek my help.

Written in 1964 by Louis Lasagna, Academic Dean of the School of Medicine at Tufts University, and used in many medical schools today.

Help is defined by those that need it; not by those that give it. - June Callwood



Welcome to Our Home

By Maxine And Brad Fisher

Dear Home Care Professional,

Welcome to our home.

Since you are new to the team of professionals who care for our daughter, I'd like to share some background with you as a way to begin our journey together.

Seventeen months ago when my wife, Maxine, and I decided to become parents again, we didn't plan to meet you. Back then, we didn't know anything about pulse oximeters, wheelchairs, bi-pap (two-way positive airway pressure) machines, feeding tubes, nebulizers, physio therapy or occupational therapy. We didn't plan for the parade of nurses, social workers, occupational and physical therapists, assistive technology specialists, medical supply personnel, respite workers and case managers who come and go. We didn't anticipate the need for 5 different doctors to monitor our daughter's progress at the local hospital and B.C. children's hospital.

Most of all, when we decided to become parents, we didn't anticipate that very bad day when a doctor told us our four-month-old baby had a year to live.

So I hope you don't take it personally when we say we have mixed feelings about your presence in our lives. Frankly, we wish we didn't need you. We wish our daughter were going to kindergym with all the other kids her age, scraping her knees, playing in her jolly jumper, being able to be left in a play pen by herself, and play with her older brother Sam. We wish we were spending our weekends playing with our kids like families with healthy toddlers and babies.



But Shira isn't going to kindergym or preschool, and she will never figure skate. Shira can't sit up, walk, or talk very clearly. She can't move her arms or legs. She will never dress or feed herself. She will always use diapers. She can't eat solid foods. She needs constant supervision and attention. She's medically fragile. Her care is physically and emotionally demanding.

We wish we didn't need you, but we do need you.

We need your positive attitude and your confidence. We need your sensitivity and patience. We need your knowledge, experience, and skills. And we need you to help us have hope, for our daughter and our family.

When you begin your work with our daughter, please carry yourself with confidence. You've may have been trained to take care for medically fragile children and now after spending 50 days in Victoria General Hospitals Pediatric Intensive Care Unit so have we. Shira's care requires remembering little things about her needs: what's safe to feed her, when and how to feed her, how to lift her and place her into comfortable positions, when to call Brad or Maxine to remove her secretions via suction, how to set up Shira's arm supports so she can play with toys, which way to turn her head when she lays on her stomach, back, sides, and watches her favourite video or looks outside.

Although none of this is rocket science, the sheer number of details can feel overwhelming to a newcomer. But if you feel overwhelmed, try not to show it. Act confident. Take initiative. Have a positive attitude but most of all ask questions and watch us as we demonstrate how Shira's equipment works. Remember there are no stupid questions and do not feel silly by asking the same question 15 times. We would prefer you understand and know what you are doing after all you are caring for our child and a human being! Write things down if it helps you. Enjoy yourself when you're working. But above all, carry yourself with confidence. Shira will feel safe if you feel safe.

When you begin your work with our daughter, please be patient with us. We may insist about certain aspects of Shira's care that don't make perfect sense to you. It may be awhile before we leave you alone with our daughter even though you're perfectly capable of taking care of her. You may wonder why we ask you to wash your hands when you enter our home and about your own health every time you sneeze or cough in our house. We realize we worry about Shira just about all the time. Please understand that we simply can't help it. From the moment the doctor sat us down and told us our baby had an incurable, untreatable disease, things were never quite the same again. We really are doing the best we can.

When you begin your work with our daughter, please be sensitive to our family's need for privacy one minute and our need for your active involvement the next minute. (As a wise home care administrator I know tells each of her new clients, "The good news is...you'll now be getting nursing and respite help in your home. The bad news is...you'll now be getting nursing and respite help in your home."). We'll try to be sensitive to the needs of your family, too. Let us know with enough advanced warning if you need flexibility with your work schedule because your mother is visiting from out of town or you'd like to attend your daughter's school play etc. It feels good for us to help someone else's family once in a while. We'll help you out whenever we can. **Please note that when it comes to Shira's Health, questions about Shira's health and Shira's well being we can be interrupted at any time no matter what we are doing. Shira's safety, and health takes precedence over everything. If we are asleep bang on the door, if we are on the phone interrupt, if we are in the bathroom bang on the door. Again, nothing is more important than Shira's immediate health and must take precedence over anything and everything with immediate prompt action.**



When you begin your work with our daughter, please take the time to get to know her as a person. Yes, you're here because of all the things Shira can't do by herself. But there's a lot she can do, too. You'll be surprised the first time you see Shira take a bath, smile and reach for your hand. Enjoy Shira enjoying the company of her older brother Sammy. Soak up her positive attitude, determination, and her smile. We think Sweet Pea is quite a girl. We hope you will, too. And when you show a genuine interest in Shira, she will fall in love with you very quickly.

When you begin your work with our daughter, please teach us what you know about caring for her. After Shira's diagnosis, one of the first things we heard from medical professionals was, "You will soon be the experts on Spinal Muscular Atrophy (SMA) and caring for Shira." But I didn't want to believe our baby had such a terrible disease, much less become an expert on it. And more importantly, if Shira really did have SMA and would die because she couldn't breathe, how in the world could we become experts on how to help her? After all, isn't that what doctors and nurses are for?

We have fast tracked on becoming experts on SMA because Shira is our daughter and we had to do whatever we could to help her. But I feel relieved every time we meet a professional (believe me these experts are few and far between) who knows more about caring for a medically-fragile child than we know, or at the very least can teach us something new. As you begin working with our daughter, tell us what you know, tell us what you've learned, tell us what you've seen. We take care of only one medically fragile child. You may have taken care of many more than one. If you have experience in providing care for the extreme medically fragile you might be able to provide us with some perspective. You can give us a reality check when we need one. You can help us take care of Shira when she's sick as her care is exponentially increased. We need your expertise. That's why you're here. Please don't hold back.



Finally, and perhaps most importantly, when you begin your work with our daughter, please help us to have hope. When we first learned Shira was affected by Spinal Muscular Atrophy, hope was a scarce commodity in our home. We didn't get any information from the doctors about living with SMA from day to day – they didn't tell us about the good days we would have together as a family in fact almost everyone we have met in the medical profession has no experience to limited experience with this disorder. Looking back, I now realize it was support groups such as www.smasupport.com and Shira's job to teach us these important lessons. After all, doctors at Victoria General Hospital and Vancouver children's hospital don't research enough or have enough time to spend with SMA kids when they are well. Parents do that and share this information through support groups such as www.smasupport.com and Dr.'s that specialize in SMA.

Our daughter is intelligent and sensitive; she observes all of our words, actions, and signals. Shira needs us to view her disability and her future with a sense of hope, whether life feels manageable at the time or not. "Shira's a weak girl," the doctors told us, quite matter-of-factly.

Indeed, Shira is a fighter. All of the children you care for are fighters. But our children can not fight without hope, specialized medical protocols and equipment. It is up to us – parents, professionals and Hashem to keep her alive!!

We appreciate everything you do!

Welcome to our home.

Sincerely,
Maxine Fisher B.A., M.Ed., B.A. Music Therapist
Brad Fisher
Sammy Fisher 4 years old
Shira Fisher (SMA Type 1 DOB 06/16/05)





"The phrase is apt to cause disquiet. There have been those among us who have arrogantly judged, from a vantage point of power, the value of a human life. They have made decisions based on their assessment of a person's quality of life about providing supports to sustain that life. This attitude peaked in Nazi Germany, where such decisions were used as the basis for genocide. We like to think that we have moved well beyond this perspective, but important decisions about people's lives are still being made from positions of power. Such practice is difficult to combat, especially in a period when responsibility of government in the area of human and environmental services is being cut back."- Introduction by J. David Baker (Quality of Life in Health Promotion and Rehabilitation)

RESEARCH

NIV Protocol Info www.doctorbach.com www.smasupport.com
Quality of Life Research www.utoronto.ca/qol/
www.fsma.org
www.planetsma.com

DOCTOR BACH IS MY HERO. I KNOW EMPERICALLY THAT DR. BACH'S NIV PROTOCOL SAVED SHIRA FROM AN UNTIMELY DEATH; .-BRAD FISHER (SHIRA'S DAD)

DR. JOHN BACH IS MY HERO

WWW.DOCTORBACH.COM (A web site created by one of his patients. A wonderful tribute to a courageous man.)

Giving Breaths, Saving Lives



The Center, which was established in 1992, is dedicated to the care of patients with neuromuscular disease and respiratory impairment from any cause. On staff are physicians from numerous specialties, including neurosciences, anesthesiology, surgery, and orthopedics, as well as respiratory therapists and speech-language pathologists. Some 60 percent of the Center's patients are children under the age of 18.

Bach contends that the majority of people with neuromuscular disorders are not managed optimally by physicians. While orthopedic problems are approached assertively, prevention of respiratory problems is given short shrift or ignored. "Families of people with neuromuscular disorders are usually told by doctors that they have three options," he says. "The first is to do nothing, or 'let nature take its course.' This ultimately results in death from respiratory failure. The second option is a tracheostomy, which makes a person completely dependent on a respirator and unable to speak. The third option, which is ours, is using noninvasive respiratory aids to facilitate breathing and coughing."

Using this third option, respiratory complications can be minimized and in many cases, avoided entirely, says the physician. His protocol utilizes close monitoring of oxygen and respiratory muscle function, non-invasive ventilation by nose or mouth as needed, and assisted coughing when necessary.

What is in Bach's powerful arsenal? A variety of devices and aids that are conceptually fairly simple. First, portable instruments such as spirometers and oximeters painlessly measure oxygen and respiration levels. When they are low, inspiratory muscle aids are used to enhance breathing. Air is administered through nose or mouth pieces or masks that are attached to ventilators. The procedure is called intermittent positive pressure ventilation, or IPPV. It is regular air, not pure oxygen, which can be harmful to those with muscle weakness (see box, above). Some people require supplemental air all the time, while others use it as needed--during a cold, for instance. Virtually everyone with neuromuscular disorders eventually requires aid for nighttime breathing, because respiratory levels drop even lower when a person is asleep.

Expiratory muscle aids involve use of manually and mechanically assisted coughing to eliminate airway secretions. The In-exsufflator, or CoughAssist, pulls air and mucus out of the lungs by applying positive pressure to the airway, and then rapidly shifting to negative pressure. This rapid shift of pressure produces a simulated cough. Bach says that even very young children can learn to cough with assistance. "Many people with DMD, SMA, ALS, and similar conditions should never require hospitalizations for breathing difficulties and can live decades longer than most people think," says Bach. Unfortunately, too many physicians think these disorders are untreatable



Quality of Life Issues

Many people unfamiliar with these diseases may think that being wheelchair-bound and dependent on these respiratory aids offer a poor quality of life. Bach disagrees. "I treat a lot of children," he says. "Their parents love them as much as any parent loves a child. They should be the judges of their quality of life, not other people." With the help of computers and technology, his patients attend school, dress up for Halloween, surf the Web and hold jobs. One of Bach's patients with ALS is a municipal court judge who has been using a computer-activated communication system as well as an inspiratory muscle aid to facilitate breathing for 15 years.

An observer may wonder if Kathleen Piazza holds a mechanical engineering degree, so familiar is she with her son's equipment. It only seems that way. Respiratory therapists from the Center, like Brian Weaver, RRT, and Louis Saporito, RRT, train family members in the use of non-invasive ventilation alternatives. Saporito has been a member of Bach's team for 18 years, and they know each other so well that they finish each other's sentences. Saporito makes house calls, working with patients and families in their homes. "The equipment can be

The house calls are not always local. Few physicians do what Bach does. So patients come from great distances for treatment and training. When they return home, they often need help in setting up in-home care. A few years ago, a set of twins from Georgia, both with SMA, came to the Center for treatment. When they flew back to Georgia, Brian Weaver went with them to help them set up their equipment at home. He also visited other healthcare personnel in the use of inspiratory aids. "Now the twins can get the care they need right in their hometown," he says. "And the protocol is in place to help other patients in Georgia."

It is the physician's goal to spread the word to all patients with neuromuscular disorders and their caregivers: *"People can survive with neuromuscular disease, and they can live meaningful lives."*

Unfortunately Shira has a terminal genetic disorder. The statistics point out that Shira has a 30% chance of living past the age of 1 and a 15% chance of living past the age of 2. We will fight the good fight in the hope that Shira lives long enough to be cured by science or well enough to live a happy life full of love.

Figures often beguile me, particularly when I have the arranging of them myself; in which case the remark attributed to Disraeli would often apply with justice and force: "There are three kinds of lies: lies, damned lies and statistics."

- Autobiography of Mark Twain

The Hazards of Oxygen

One of the most common treatment errors for people with neuromuscular disorders is oxygen therapy, which Bach likens to "putting a Band-Aid on a cancer." He says that oxygen should never be used for people with neuromuscular conditions unless pneumonia or RSV has resulted in the need for intubation, intensive care and resuscitation.

Oxygen turns off the brain's drive to breathe and greatly increases the likelihood of ventilatory failure, basically allowing the blood's carbon dioxide to increase to levels that render a person unconscious and cause him to stop breathing entirely (respiratory arrest).

Usually decreases in blood oxygen levels are caused by airway secretions that the patient is not strong enough to clear without training in Bach's methods and the secretions themselves can also result in respiratory arrest.

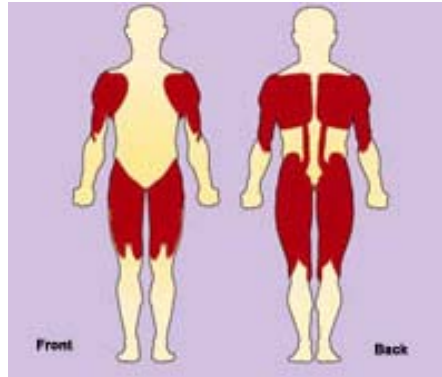
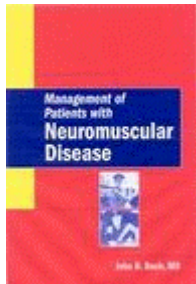
A Recent Study

How effective is Bach's method in reducing hospitalization of children with SMA?

A long-term study by the physician examines 52 children with SMA who were treated at the Center. The children in the study had an average of 1.53 hospitalizations per year by their third birthday. Between the ages of 3 and 5, the rate dropped to 0.33 hospitalizations a year, and after the fifth birthday, dropped even further to an average of only one hospitalization in more than 20 years.

The results of the study will be published in an upcoming issue of Pediatric Pulmonology

I HAVE BOTH OF THESE BOOKS IN SHIRA'S BEDROOM ON HER BOOK SHELF. DR. BACH'S PROTOCOL FOR NON MECHANICAL VENTILATION AND MANAGEMENT OF PATIENTS WITH NEUROMUSCULAR DISEASE



THE ROAD NOT TAKEN

Two roads diverged in a yellow wood,
And sorry I could not travel both
And be one traveler, long I stood
And looked down one as far as I could
To where it bent in the undergrowth;
Then took the other, as just as fair,
And having perhaps the better claim,
Because it was grassy and wanted wear;
Though as for that the passing there
Had worn them really about the same,
And both that morning equally lay
In leaves no step had trodden black.
Oh, I kept the first for another day!
Yet knowing how way leads on to way,
I doubted if I should ever come back.
I shall be telling this with a sigh
Somewhere ages and ages hence:
Two roads diverged in a wood, and I-
I took the one less traveled by,
And that has made all the difference.

-Robert Frost

SHIRA FISHER TREATMENT SCHEDULE

12 AM – FOOD FINISHED

1. Vent Line/Flush Line 10ML of water
2. Change Diaper
3. Apply zinc oxide karaya powder (If Needed)
4. Move oximeter sensor
6. Check BiPAP for alignment and Leak

2AM - FOOD

- 220 ML of Food
- Change Diaper
- Apply Zinc Oxide Karaya Powder (If Needed)
- Move sensor
- Check BiPAP for alignment and Leak

4AM – FOOD FINISHED

1. Vent Line/Flush with 10ml of water
2. Move oximeter sensor
3. Change diaper
4. Zinc Oxide Karaya Powder (If Needed)
5. Check BiPAP for alignment and Leak

6AM - FOOD

1. 220 ML of Food
2. 3 mls of liquid vitamins in food
3. Diaper Change
4. Zinc Oxide Karaya Powder (If needed)
5. Check BiPAP for alignment and Leak

8AM- FOOD FINISHED

1. Vent Line/0.5 mls Ranitidine/Flush 10mL 02
2. Remove BiPAP
3. Disassemble Food Pump/clean bottle

When Shira Wakes:

Chest Physio Therapy

- A) percussions - upper loabs-both sides-back both lobes
- B) cough assist - Settings:inhale 1.5, exhale 1, Pause 1
- C) inhale pressure 40, exhale pressure 40
5 rounds of 5(5breaths with no pause)
- D.) Suction: Suction between each set or when you see bubbles or the need to suction
- E.) Move oximeter sensor
- F) Empty suction
- G) Record Treatment In Book

Physio Therapy see pages 16-18 after physio session is over place Shira in her AFO's and Wrist orthotics. Take her out of her bedroom to the living room for play time.

MEDICATIONS

- **Ranitidine** – 0.5 mls 8am and 8pm through g-tube
- Vitamins**-3ml 10pm feeding in food

If Shira is in need of other medications a directive will be set up for that day. Do not give Shira any other meds other than what is outlined in this directive without the consent of either Maxine or Brad. Also, all treatments are to be recorded in the daily journal.

DIAPER CHANGES

- Every 2 hrs. or as needed

Coat buttocks and Labia with with Zinc Oxide Karaya powder ointment if there is a rash.

NECK

- Make sure neck is dry

- If rash is present clean using soap and water

Apply Viaderm-K.C. Ointment

SHIRA'S PROTOCOL

PLEASE NOTE THAT THIS PROTOCOL WAS DEVELOPED SPECIFICLY FOR SHIRA'S NEEDS AND SHOULD BE USED AS A GUIDELINE FOR DEVELOPING A PROTOCOL FOR YOUR CHILD. EVERY CHILD IS DIFFERENT!

SCHEDULE CONTINUED ON THE NEXT PAGE

10AM - FOOD

1. 220 ML of Food
2. 2 capsules of Acidopholous
3. Diaper change,
4. Zinc Oxide Karaya Powder (If Needed)
5. Observe Shira for signs that she may need to be on BiPAP
6. Record Oxymeter read out

12PM – FOOD FINISHED

1. Remove food line/Vent Line/
2. Flush with 10ml of water
3. Move oximeter sensor
4. Change diaper, Zinc Karaya Cream (If Needed)
5. Check bedding- change as needed- t shirt
6. If Shira is not sleeping take her around the house in the stroller - attach oximeter

2PM - FOOD

1. Acidopholous
2. 220 ml of Food
3. Diaper Change, Zinc Oxide Karaya Cream
4. Bipap break and nap
5. Make sure bed is dry and clean

4PM – FOOD FINISHED

1. Shira wakes up let her linger on bipap for about 20 minutes or until she asks to be taken off bipap
2. Flush with 10ml of water
3. Diaper, Zinc Oxide Karaya Powder (If Needed)
4. Place pump in charger
5. Wash feed bag and drain

6PM – FOOD

1. 220 ml of Food
2. Diaper, Zinc Oxide Karaya Cream (If Needed)
3. Oxymeter

8PM – FOOD FINISHED

1. Remove Food Line/Vent Line/Clean bottle and line
2. Replace Pump back on charger
3. Wait 15 minutes after food finishes to give Shira CPT

Chest Physio Therapy:

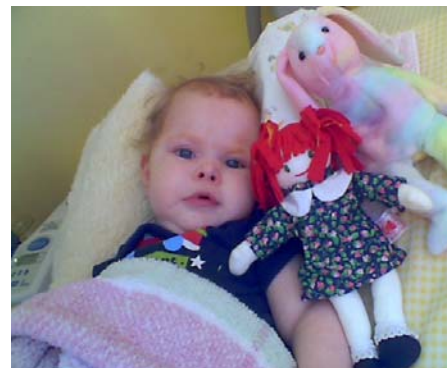
- A) percussions - upper loabs-both sides-back both lobes 10 minutes
- B) cough assist - Settings:inhale 1.5, exhale 1.5, pause 1
inhale pressure 40, exhale pressure 40
5 rounds of 5 breaths (Suction between each set or when you see bubbles etc.)
- C) 1.33ml of ranitidine through g-tube
- D) Give Meds if needed - Change Diaper- T shirt
- E) Move oximeter sensor
- F) Empty suction
- G) Phisyio Therapy see pages 16-18
- H) **Record Treatment In Book**

10PM - FOOD

- 2 Capsules Acidopholous
- 220ml of Food
- Change diaper (Zinc Oxide Karaya Powder if needed)
- Move Sensor

BATH TIME

- Shira likes a warm bath
- Special bath seat sits in tub
- Wash creases i.e. neck, hands, behind knees etc. thoroughly to avoid rash
- Wash Hair
- Dry Shira Thoroughly
- Apply Zinc Oxide Karaya Cream to buttocks and labia if there is a rash
- Neck-Apply Viaderm –K.C. Ointment
- G-Tube-Consult Brad or Maxine



Night Respite Protocol next 3 pages

DESCRIPTION

Night care is as important as day care! Because Shira can not move it is imperative that Shira is rotated often to maintain circulation and ward off bed soars. Night respite gives us parents a time to sleep so that we can better care for Shira during the day insuring a high quality of life is maintained. Please note that at no time are you required to perform any medical interventions. Your main duties are to insure Shira's comfort, feed Shira, monitor Shira throughout the night and inform us parents if there is a problem. Please be as quite as possible during the night as we are sleeping in the rooms right next door to Shira.

MAIN DUTIES

- Turning Shira – Positioning
- Feeding Shira – Kangaroo food pump system
- Mask alignment
- Diaper Changes
- Oxymeter Sensor placement
- Recording

WHEN YOU ARRIVE

1. Door will be open
2. Wash hands with hand sanitizer at door
3. Lock door
4. Wash hands in sink upstairs with soap and water before entering Shira's room sing happy birthday to yourself while washing with hot soapy water
5. Dry with a paper towel not a cloth towel
6. Leave any coats, bags etc. which are used out in the general public outside of Shira's room.
7. Shira is usually sleeping or is ready to sleep when you arrive

TURNING SHIRA

1. Turn Shira onto her back
2. Quick mask adjustment – make sure nostrils are not covered by mask
3. Turn Shira onto her side
4. Immediately straighten hose and make sure it is aligned over the center of her head between her eyes
5. Tilt head so that mask is not touching bed making the mask go out of alignment
6. Make sure Shira's head is in alignment with her back. Shira should not have her chin buried in her chest or her chin pointing too far away from her chest
7. Place bunny against Shira's soother
8. Pull arm against bed out from under Shira so that she is not laying on her arm

Place the 3 receiving blankets in a step like fashion over arm on bed all the way over the bunny

Lift Shira's bottom and re align shirt

Place pillow between legs

Legs should be in fetal position.

IMPORTANT – Look at Shira. If you couldn't sleep in the position you left her in Shira probably can't either!

NIGHT PROTOCOL CONT'D

OXYMETER

- Change over every 2 hours
- Silence Alarm
- Change over sensor
- Never leave the room until you see Shira's SPO2 numbers reading on the display

CHANGING SHIRA'S DIAPER

- Turn Shira onto her back
- Turn Shira's head toward direction of final destination and place a receiving blanket under Shira's face like a pillow
- Adjust mask
- Place bunny over soother
- Make sure legs are positioned and supported
- Open new diaper on bed
- Release soiled diaper and clean Shira over her soiled diaper. Lift Shira by holding her ankles in your hand. Never clean Shira over the bed or lay her on the bed without diaper as accidents do happen.
- Place Shira on her new diaper and close the diaper around Shira precisely but not tight as Shira is a stomach breather and we do not want to impede her breathing.
- Remove the receiving blanket under Shira's face and proceed to turn Shira to her final destination.
- Proceed to #3 under the heading "TURNING SHIRA."

FEEDING SHIRA

CLEANING CONTAINER

1. Make sure line is open
2. Use hot water and fill container then shake out excess food. Do this procedure 2 times
3. Fill container with hot water and hang on hook to drain
4. If line has food in it and water is not flowing squeeze the cylinder until water pushes food from line and water flows
5. Shake cylinder to clean container
6. Let water run through entire line until container is empty
7. If container smells use bottle brush and a tiny amount of soap and clean container
8. Make sure all soap is thoroughly rinsed from bottle

FILLING CONTAINER WITH FOOD

1. Shira's food is kept on top right shelf of refrigerator and no where else!!!
2. Shake bottle well mixing food
3. Fill stainless steal bowl with hot water
4. Fill baby bottle with 220mls of food and place in hot water for 5 minutes
5. Turn off line by crimping the line
6. Fill bottle with food and place lid on bottle then hang on hook
7. Hold cylinder in left hand and control flow with right hand
8. Don't overfill cylinder!! Make sure liquid does not surpass nipple in cylinder
9. Run food through line and turn off before food exits line.
10. Rinse line under hot water and wipe with paper towel
11. Place in feeding machine.

MOST COMMON REASONS FEEDING MACHINE ERRORS

- 1) Line crimp on g-tube or container line are crimping the line closed
- 2) Cylinder is too full
- 3) g-tube line is crimped during a turn or obstructed by shira lying on it
- 4) line is crimped or obstructed on machine side

NIGHT PROTOCOL CON'T

MASK ADJUSTMENT

Mask adjustment is the most difficult part of the night job. I ask that you do not remove the mask from Shira's face without me present as Shira needs suctioning and attention at this point in case she panics due to decompression. The mask is a very light silicone and just sits on Shira's face as apposed to being strapped tightly to her face. This prevents soars caused by the mask and head gear and allows for a more comfortable sleep.

1. Grab the mask between your index finger and thumb on both sides of the mask and gently move the mask from side to side without lifting the mask.
2. The objective is to expose Shira's nostrils and remove any leaks from the mask
3. Check mask leak rate on BiPAP Machine by pressing down arrow 2 times and read the LR. The rate should not exceed 17 but should ultimately be at 15 or lower.
4. The most common cause of mask leak is accidental shifting of head gear and misaligned hose.
5. If the head gear becomes badly aligned while turning Shira exposing her nose you must wake me up right away to re set Shira's mask do not attempt this on your own please.

CONSOLING SHIRA

1. Shira does not like effleurance!!! Shira likes to be touched like you mean it!!
2. Pat Shira's back using the palms of both hands. Facing Shira's face put your hands over Shira i.e. near her bottom and one in the middle of her back and start to pat Shira firmly. Shira should be rocked by this motion which in turn will soothe her and help her fall asleep.
3. Rub the bunny against Shira's soother
4. Try both of the above procedures at the same time.

SUPPLIES

- Paper towels and Kleenex are stored in the broom closet directly across from bathroom door.
- feeding pump bottle and line are in Shira's cupboard.
- Diapers and wipes are in Shira's closet directly underneath the top of the shelf and stored to the left of the shelf.

POWER FAILURE

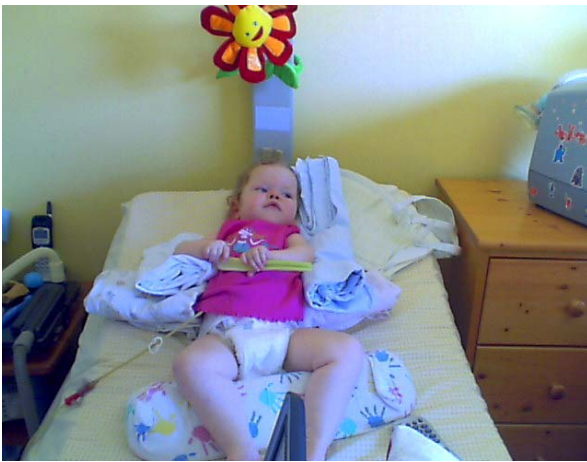
1. Immediately plug in AC adapter to bipap and turn on
2. Wake up Brad
3. Flash light on shira's med counter

WHEN YOU LEAVE

1. Open Shira's door
2. Open Brad's door
3. Turn monitor on in kitchen so that it is audible from bedrooms
4. Turn off light and turn off heat in Shira's room
5. Make sure you have all of your belongings before exiting the house
6. Lock lower lock on front door and shut door behind yourself.

POSITIONING A MAJOR CAUSE FOR DESAT AND CHOKING

Positioning is one of the most important aspects of Shira's care. SMA Type 1 children have major problems managing their secretions. When Shira is placed in any position other than flat she must be continually monitored in case she needs suctioning or DESATS. Shiras favourite positions are 1) Flat 2) 25 degrees on bed and wedge for the floor 3) 35 degrees 4) sitting up. Shira is a stomach breather and sitting in an upright position in the tumble form chair restricts her breathing constant monitoring is a must. **When in any position other than flat (and connected to BiPAP and Oxymeter) it is imperative that Shira be on Oxymeter and be continually monitored 24/7!!!**



Position #1

When Shira is in her bed it is important to support her legs to prevent hip displacement. Shira needs supports under her shoulders to her elbows to allow Shira to move her arms in front of her and her head must be supported towards the angle she is facing. Shira must be on Oxymeter!



Position #2

When Shira is on the floor or quarantined downstairs (during family illness using the crib) we use the 25 degree wedge. If Shira is not on Bipap she is continuously visually monitored. Please suction Shira as much as needed as she can not manage her secretions on her own. Oxymeter!!



Position #3

Tumble Form chairs help Shira get used to sitting up. Because SMA children are stomach breathers sitting up restricts their breathing and causes them to desat after a period of time. Never leave Shira in this seat un attended and without her oxymeter attached. Suction Shira as much as needed. If Shira is unable to be in a sitting position in the tumble form remove it from the base and utilize the chair without the base and adjust Shira to an appropriate angel.



SHIRA MUST ALWAYS BE ATTACHED TO THE OXYMETER. SMA KIDS PLUG, HAVE VASO VAGAL RESPONSES, EXTREME BRADY ETC. PLEASE VISUALLY MONITOR SHIRA RELENTLESSLY WHEN NOT ON BIPAP AND SITTING UP

CAR TRAVEL



Physiotherapy Range of Motion Suggestions

Next 3 pages

All range of motion with Shira must be done very slowly and carefully. Children with SMA are known to often have fragile bones that fracture easily and there is almost no muscle support at her joints so subluxation or dislocation would be very easy.

The following are the range of motion activities for those muscle groups where Shira has or has had some tightness. For all of these movements, I support the limb with two hands and only move as far as is comfortable, very slowly. When I encounter mild resistance or if Shira furrows her brow, I just hold it there, perhaps “wobbling” a little from side to side and then move further only if I feel relaxation. I prefer to do one slow stretch of one to two minutes rather than many fast movements. Shira likes to help with these activities and can assist toes up and down, arms up and down and all hand movements

It is fine to do the general mobility exercises: e.g. the baby Yoga that Brad has shown me or bicycling movements as long as the above principals are kept in mind. Pay special care when rolling over, as it is very easy for her to get arms in a position where shoulder subluxation could happen



Support Shira at upper arm and at hand/wrist. Ease her arm forward and up towards her ear. When you feel slight tension (or see Shira frown) DO NOT FORCE the arm further. Just gently shift it, with tiny side-to-side movements, until it relaxes. Then you may be able to ease the arm a little further.

Make sure that Shira's head spend equal time turned to either side. If it is too hard to move the TV or her respiratory equipment, change Shira so that her head is at the foot of her crib.



There is slight tension on the “little finger side of Shira's wrist. Gently bring hand in line with lower arm, and then ease the web space of her thumb open. Do not pull on the end of her thumb. Shira has a tendency to twist her forearm so that thumb is down. Gently untwist forearm to bring thumb up.



Gently open fingers and straighten wrist. I often spend some time doing gentle but deep massage of her palm, as a lot of the small muscles feel tense. She enjoys "round and round the garden" during this stretch.



Ease Shira's head and shoulders to one side, while her pelvis stays flat. This provides elongation along her thorax and pelvis. Change sides.



Roll Shira's pelvis to one side while her shoulder stays flat. Change sides



Ease Shira's foot up towards a right angle. Never force- she can be quite sensitive around her feet.



I start with the knee bent and gradually straighten the knee. This one has been quite tight, left foot more than right, so I will often spend 10 minutes or more on this one.



Once she is comfortable with the knee straight, I let go of the foot and raise the whole straight leg very



All the little muscles in Shira's foot get very tense. She has a strong tendency to curl toes. Gently uncurl toes. First do one at a time. Once they relax you can uncurl them all.



Slow deep massage of the long muscles in the sole of the foot helps to relax some of the stiffness within the foot so that it allows more very gently twisting (big toe up, big toe down)

COUGH ASSIST PRCEEDURE

BRIAN'S RESPONSE TO SHIRA'S PROTOCOL

Brian X Weaver MS, RRT-NPS, RPFT-Adult and Neo/Ped Clinical Specialist
Respiratory Therapy University Hospital / UMDNJ, Newark, N.J.
Email: weaverbx@umdnj.edu Phone: (973)972-0700

The cougholator (now the cough assist) shows In 1.5 out 1.5 pause 0.5. I do not use any pause at all. The more pause, the less secretions I have been able to get moving aggressively. As the kids get older they learn how to better utilize the machine. The cough assist is also lung therapy as it expands the upper lobes and chest cavity and is the most important therapy in keeping SMA kids healthy. In your cold section your adjustment is to frequency which is good but your change to procedure is also to shorten In and Expiratory times and this is not good in fact it is opposite of the best adjustment (shorter pause time also, I would not even use a pause or a pause of 1 max). I prefer to do it manually if possible. The sets should be increased to 3, 4 or 5 consecutive breaths each and do 3, 4 or 5 sets total with the in and out time the same as always or a little longer if tolerated and keeping the mask on for consecutive breaths during each set with a rest between sets.

The biggest point I can shout and I cannot shout this loud enough is not to apply this to other kids. They can look at this protocol and see if there is anything they can use but this plan you wrote is for Shira and only for her! This plan may work well for her but may be a disaster for others. Each child must have their individual plan that caters to their needs. Your plan includes some basics but it also contains specifics that could very easily harm other babies. If you discuss this with other families be very clear that this is not a general care plan. Make it clear that this is what you do and they should form their own plan with their medical caregiver or the closest one of us that can review their case.

I do not post that warning lightly. We need the word to get out but I have seen horrible results when a parent (not that you would) suggests care for another based on the care that they use to their child. You a good man and father just be careful with advice. Do not stop helping others just be very careful about directions.

I will give you a simple and not serious example. Parents relay my statement in training if tolerated. This is applied to "medically tolerated." In advice from one parent to several it was changed to, "if the baby tolerated it", as in, "did not mind it." This caused babies to get less than the needed treatment because they looked unhappy. Of course when I found out I flipped. You see I care about the children and their happiness but when faced with a critical situation then I'm sorry you do not like this but tough. It is life (not liking it) or death (not doing it) and I always choose life. See my point. When we talk about the bigger things it gets worse but if you say, "this is not what you should do, this is only what I do then they must evaluate and check out how it effects their child."

SHIRA'S DIET THE AA DIET

WHAT IS THE AMINO ACID DIET?

Mary Bodzo

smbodzo@msn.com ♦ (352) 245-9119

There are links to this site and information on
www.smasupport.com

Technical Features - VIVONEX[®] Pediatric

Caloric Distribution:

Protein: 12% (Free Amino Acids)

Carbohydrate: 63% (Maltodextrin, Food Starch Modified (Corn))

Fat: 25% (MCT Oil, Soybean Oil)

Caloric Density: 0.8 cal/mL*

Nonprotein Calorie: Nitrogen Ratio: 200:1

Water Content: 893 mL/1000 mL

100% NAS/NRC RDA:** 1-6 years in 800 calories (1000 mL); 7-10 years in 930 calories (1170 mL)

Osmolality: 360 mOsm/kg water*

Lactose free, low residue

May be suitable for a gluten free diet



Many kids also use pediatric Tol-erex which contains less fat. Many of the kids start on Vivonex and switch over to the Tolerex after a year old.

Below is Shira's Diet for 1 24 hour period. The diet is given as a bolus feed 2 hrs. on and 2 hours off 6 times a day. Every child is different and it is advised that you start with a basic diet of the Vivonex or Tolerex slowly adding supplements, baby food ect. In small increments over a period of a week or two per new ingredient making sure your child can tolerate it and that secretions and heart rate remain stable. Calculating the correct calories and proteins is essential. All children are different and experimentation and observation are key.

RECIPE 1 DAY SUPPLY

1. Blend 2 packs to 700 mls of water
2. 3ml of liquid vitamins
3. 16th of a teaspoon of Glutamine
4. 100 mls of cooked sweet potatoe no skin
5. 350 mls of fresh juice
6. 100 mls of prune juice
7. 50 mls of banana
8. 40 mls of avocado

We tried baby food but Shira had a hard time digesting it so we went to fresh juice which in our opinion has a lot of nutrients and vitamins missing in processed foods.

JUICE RECIPE FOR 4 DAYS

1. Juice 2-4 bunches of spinach or Kale if no spinach available
 2. 3 celery sticks
 3. 3 large apples –6 medium apples
 4. 3 ripe large size pears
- Careful with pears as they are an excellent natural laxative if your child developes runny bowel movements try cutting back on the pear.

Strain juice through medium fine strainer.

NIV PROTOCOL EQUIPMENT

10-11 get Shira ready for bed bipap etc.



Emerson Cough Assist

The non-invasive CoughAssist safely and effectively clears retained broncho-pulmonary secretions, reducing the risk of respiratory complications. The CoughAssist achieves this by gradually applying a positive pressure to the airway, and then rapidly shifts to negative pressure. The rapid shift in pressure produces a high expiratory flow from the lungs, simulating a cough.

This technique, referred to as "mechanical insufflation-exsufflation," avoids airway damage while offering patients greater comfort and quality of life without the use of an invasive procedure. It has been proven effective for patients with an insufficient ability to cough due to Poliomyelitis, Muscular Dystrophy, Spinal Muscular Atrophy, Amyotrophic Lateral Sclerosis, Myasthenia Gravis, Guillain-Barre Syndrome, Multiple Sclerosis, or other neurological disorder with some paralysis of the respiratory muscles, such as spinal cord injury.

- **Indications for Use:** Any patient unable to cough or clear secretions effectively due to reduced peak cough expiratory flow (less than 3 liters per second), resulting from high spinal cord injuries, neuromuscular deficits or severe fatigue associated with intrinsic lung disease, is a candidate for the CoughAssist.
- **Contraindications:** Any patient with a history of bullous emphysema, known susceptibility to pneumothorax or pneumo-mediastinum, or known to have had any recent barotrauma, should be carefully considered before use.

The CoughAssist can be used with a facemask, mouthpiece or, with an adapter, to a patient's endotracheal or tracheostomy tube. This device can be used on adult and pediatric patients at home or in a hospital/institutional environment.



The BiPAP Synchrony provides more natural and comfortable noninvasive ventilation specifically for COPD patients and others suffering from respiratory insufficiency. ALS or neuromuscular patients may also benefit from the therapy provided by Synchrony. Synchrony can be adapted to meet a patient's needs by offering flexibility in treatment options. The unit can be upgraded from Spontaneous Mode (S) to a version with Spontaneous/Timed Mode (S/T). For greater flexibility, the unit has continuous positive airway pressure, Timed- and Pressure-Control Modes. Digital [Auto-Trak](#) SensitivityT has enhanced leak compensation to allow for better matching of the patient's breathing pattern. Vent Ramp and RiseTimeT also aid in the delivery of comfortable ventilation. Features a broad pressure range of 4 to 30 cm H₂O and the ability to collect patient therapy, compliance and machine data using an optional built-in modem. Traveling is easy with the Synchrony, as it is DC capable and weighs only six pounds.

NIV PROTOCOL EQUIPMENT cont'd



DEVILBISS SUCTION PUMP WITH BATTERY PACK

Features: Easy-to-read, built-in regulator and vacuum gauge for quick, accurate adjustments. High-performance capabilities, up to 550mmHg vacuum and 27 lpm flow. Convenient, 800 cc, single-patient use canister for easier cleaning and less costly replacement. Small, lightweight design. 12V DC operation with cigarette lighter cord. Also available with built-in battery for portability. Battery operates up to one hour when fully charged.



KENDALL KANGAROO PET PUMP

This pump is used to administer Shira's food through her g-tube. The pump is not gravity fed it is mechanical so the pump can be lower than the patient. The pump should always be placed on the charger between feeds. I like to leave the pump on the charger in Shira's room during feeds to insure that the pump is always fully charged.



MASSIMO OXIMETER

SHIRA USES THE MASSIMO OXIMETER TO MONITOR HER BLOOD OXYGEN SATURATION LEVELS. THIS DEVICE SHOULD BE USED IN CONJUNCTION WITH AN INDEPTH MONITORING OF THE PATIENT MANUALLY. NEVER RELY SOLELY ON THIS MACHINE AS IT IS A MACHINE. ALWAYS TRY TO FIND THE SOURCE OF WHAT IS MAKING SHIRA'S SAT LEVELS FLUCTUATE AND USE THE SAT MONITOR AS A TOOL TO INFORM YOU AND HELP IDENTIFY THAT SOMETHING MIGHT BE WRONG.

THE OXIMETER SENSOR SHOULD BE MOVED EVERY 2 HOURS OR MORE TO PREVENT THE LIGHT FROM BURNING HER FOOT.

**If you want to tell me that the stars are not words,
then stop calling them stars.**

- Jack Kerouac

BiPAP MACHINE



Shira can be awake and playful with the BiPAP on.



**Shira asleep with the BiPAP and soother.
Shira's favourite side is with her right side up.**



Bereft

Where had I heard this wind before
Change like this to a deeper roar?
What would it take my standing there for,
Holding open a restive door,
Looking down hill to a frothy shore?
Summer was past and day was past.
Somber clouds in the west were massed.
Out in the porch's sagging floor,
leaves got up in a coil and hissed,
Blindly struck at my knee and missed.
Something sinister in the tone
Told me my secret must be known:
Word I was in the house alone
Somehow must have gotten abroad,
Word I was in my life alone,
Word I had no one left but God.

-Robert Frost

BiPAP PROTOCOL

OBSERVATION AND PROBLEM SOLVING HOW TO MAKE SHIRA MORE COMFORTABLE

WHEN DOES SHIRA NEED THE BIPAP?

Shira is diagnosed as a ventilator dependent patient. Shira does not need to be on BiPAP 24 hours a day but due to Shira's lack of musculature caused by SMA she needs to be supported in her breathing so that she can recuperate her strength throughout the day and so that she may have a more restful and rejuvenating sleep at night. There are no specific times as to when Shira has to be on BiPAP; BiPAP is administered as she needs it. Below are the main symptoms that will alert you to the fact that Shira needs to be placed on the BiPAP:

- 1) Diaphoretic sweaty head to the touch
- 2) Flaring nostrils
- 3) Labored and Jerky Breathing (Remember that Shira is a stomach breather to begin with but you will see her jerk with each breath rather than breath in a gentle rhythm).
- 4) Mottled skin. You will see her veins through her skin. She will start to look like a road map on her legs, face cheeks and arms
- 5) Cranky, inconsolable, just not happy no matter what you do. Look at the Oximeter, what does it say? Does she need a suction and BiPAP?
- 6) When Sick, Teething etc.
- 7) After an emergency situation so that she can recuperate.

It is important to remember that these symptoms do not have to appear all at once. Only one might be prevalent.

WHEN SHIRA IS ON BIPAP

All masks have a natural leak rate. This is different than the masks leaking out the sides!!

1. Make sure the BiPAP machine is on before placing mask on Shira.
2. Suction Shira before placing the mask on her face.
3. Suction Shira immediately after securing the mask before final adjustment.
4. Place soother in Shira's mouth it helps her equalize pressure and direct the airflow.
5. It is of utmost importance to be diligent in preventing Shira's eyes from drying out when on the BiPAP machine. Make sure that Shira's eyes are not watering. Watering is the number one sign that the mask has a leak.
6. Mask alignment: If you try, but can not align the BiPAP mask on Shira's face due to Shira's position then remove the mask, place Shira on her back and place the mask back on Shira's face repeating steps 1 -4 again.
7. Scroll down on BiPAP machine and make sure the leak rate is 15 and under. Do not tighten mask to prevent leaks adjust the mask by moving it around on her face. A tight mask will cause sores.

BIPAP PROTOCOL CONTINUED ON NEXT PAGE

BiPAP PROTOCOL CONTINUED

OBSERVATION AND PROBLEM SOLVING HOW TO MAKE SHIRA MORE COMFORTABLE

WHEN TO REMOVE THE BIPAP

1. If you just placed the BiPAP on Shira's face and she is continually crying even after placing the soother in her mouth, checking for leaks, making sure Shira is in a comfortable position; there is a good chance that Shira just does not feel that she needs to be on the BiPAP. Remove it!
2. When Shira wakes up in the morning or from a nap she likes to have the BiPAP removed immediately.
3. Wash Shira's face with a warm wash cloth.
- 4. BIPAP REMOVAL: When you remove the BiPAP place Shira on her back, completely remove side straps, lift up the mask over her head, raise her head and gently remove the head gear making sure the side straps do not catch Shira's ears. Do not pull the head gear against her skin as you may scratch Shira with the Velcro end straps.**

She does not know
In the softest of ways.

In the way of a child who
Does not understand diamonds,
She has not found
That aperture of light.

Has not learned to read the looks
Or catch her truth in mirrors -
In the playings of light,
She stands beside herself.

In the most innocent of ways
So quiet she's all there is,
Has not recognized
That her eyes look like soul.

In the barefoot path
Of an uncrowned queen,
In the way a gentle breeze
Only whispers the storm.

In the softest of ways
She does not yet know,
That she is beautiful

- Grant van der Vijver, 2001



BED POSITIONING

SHIRA'S FAVOURITE

When positioning Shira think about how you would like to lay on your bed. Shira needs support. Make sure that her upper shoulder is supported by her arm being supported with 3 receiving blankets. Shira also likes the lambs wool weighted bag at her backside and her legs separated by a pillow.

This system is used whether Shira is on her right side or her left side.

Also make sure that Shira is placed high up on the bed to so that her chin and head remain in alignment and are not forced to either side blocking her airway



3 RECEIVING BLANKETS



WEIGHTED LAMBS WOOL BAG

IMPORTANT

IT IS IMPORTANT TO NOTE THAT MANY OF THESE KIDS INCLUDING SHIRA CAN NOT REST AGAINST A 30 DEGREE WEDGE OR SIT IN AN UPRIGHT POSITION FOR LONG PERIODS OF TIME AS THEY DESAT. MANY OF THESE KIDS ARE MOST COMFORTABLE FLAT OR UP TO 20 DEGREES. SHIRA SLEEPS AT ABOUT 15-20 DEGREES.

FLOOR POSITIONING AT PLAY STATION



Shira is positioned on the floor on top of a blue workout mat and a wool fleece.
Shira's legs are supported by a triangular foam support. It is important to support Shira's legs and try and prevent hip dislocation and stress injuries. Make sure Shira's neck and head is supported by a folded receiving blanket on each side of her head.

Place Shira on the floor and then place the play station over top of Shira



The toys on the play station are held in place with Velcro straps. To keep Shira's interest make sure that you move the toys around. Also the mirror can be placed directly above Shira or on the sides.

Shira loves to look at herself in the mirror.

Unlike these photos Shira's head should be supported on both sides by receiving blankets.



Receiving blankets should also be placed beneath Shira's shoulders. This can be done by rolling up a receiving blanket or folding a blanket. Raising Shira's arms off the ground by an inch gives her more mobility and gives her more movement.

SHIRA FISHER'S EXTREME BRADY PROTOCOL

AMBULANCE 911

ADDRESS: 4478 MAJESTIC DR. NEAR ASH AND MAJESTIC.

PICU Dr. Amanda Barclay and Dr. Raphael Beck @ PICU 727-4186 Let them know we are on the way to emergency at VGH.

EXTREME BRADY PROTOCOL (DESATURATION)

- 1.) If oximeter alarms examine Shira for signs of a plug causing extreme Brady. Look at her face is she struggling?
- 2.) Suction Shira to try and remove any upper airway secretions.
- 3.) If Shira's colour is good use cough assist: Inhale Pressur40—Exhale Pressure 40 then make sure Inhale is set at 1.5 seconds and exhale is set at 1.5, pause 1. Give Shira 5 rounds of 5 with suction in between each round.
- 4.) Is Shira breathing on her own? Look at colour!
- 5.) If Shira is still struggling for breadth but colour is good give her chest physio 5-10 percussions on each upper lobe and sides. If Shira is still struggling but colour is still good not a dusky grey blue then give another round of cough assist and then suction.
- 6.) If Shira's colour is turning a dusky blue grey bag her immediately for 5-10 breaths giving her a break.
- 7.) Re evaluate—Colour and is Shira breathing on her own—Sat levels on oximeter
- 8.) If Shira is able to breath on her own and colour is good place BiPAP on Shira and suction then let her rest.
- 9.) If Shira is able to breath on her own but SAT levels O₂ remains below <90 O₂ attach oxygen and increase litres as needed until Shira reaches <94 O₂. then call an ambulance and phone PICU at 727-4186 let them know we are bringing Shira in to emergency and to call Dr. Beck.or Barclay whoever is on call.
- 10.) If Shira can not breath on her own keep bagging Shira with supplemental O₂, call the ambulance and Dr. Beck and Dr. Barclay at PICU and let them know we are on the way 727-4186

TELL AMBULANCE THAT SHIRA MUST BE ADMITTED TO EMERGENCY FIRST UNLESS OTHERWISE AUTHORIZED BY THE PICU TO ADMIT HER DIRECTLY TO THE PICU.

PEDIATRIC AMBU BAG

1. KEPT IN CABINET BELOW OXIMETER

OXYGEN

1. KEPT TO THE LEFT OF EQUIPMENT SHELF ON THE FLOOR
2. WRENCH IN POUCH 1/4 TURN TO OPEN VALVE.
3. LITRES OF OXYGEN ADJUSTED ON TOP OF REGULATOR AT THE END OF CYLANDER.

OXYGEN ADAPTER FOR BIPAP

1. HOSE STORED WITH OXYGEN TANK
2. ADAPTER IN HOSE.
3. DISCONNECT BIPAP CLEAR WHITE HOSE AND GREY HOSE AND INSERT ADAPTER TO GREY HOSE AND SMALL 6 INCH ADAPTER HOSE TO CLEAR PLASTIC BIPAP HOSE.

SHIRA'S PROTOCOL DURING A COLD

These steps should be done in the order shown below every 4 hours while Shira has a cold.

- 1.) Chest physio therapy (see next page) every 2-4 hours depending on how sever cold is.
- 2.) Cough assist machine for 5 sets of 3-5 breaths no pause
*inhale pressure 40/exhale pressure 40, inhale 1.5 seconds, exhale 1.5 seconds, pause 0 or 1
- 3.) 2ml of infant Motrin every 6 hours and 1.75ml of infant Tylenol every 3 hours.
- 4.) Flovent X 1 shots 8 a.m. and 8 p.m. administer via aerochamber
- 5.) Ventolin X2 every 4 hours. Administer via aerochamber unless Shira has elevated heart rate.
- 6.) Check for Constipation

***RECORD ALL
TREATMENTS***

TEMPERATURE READING

Take Shiras temperature often every hour or less.

COUGH ASSIST

Use the cough assist every time Shira sounds rattley or has trouble coughing secretions. You can't use this machine too much. If Shira can't get mucous out of the back of her throat, suction her through the mouth with a #10 French catheter.

If Shira is extremely congested you can go down her nose using an #8 French catheter. Be sure to lightly lubricate the catheter with ky jelly.

BIPAP

Use the BiPAP machine every time Shira goes to sleep. Also use the machine when Shira takes naps. Shira is even weaker during colds.

OXIMETER

Use the oximeter to check Shira's oxygen saturations throughout the day. If Shira's oxygen saturation level is less than 94% use the cough assist to clear secretions and then recheck the oximetr. If the oxygen saturation is 92% or less and the cough machine is not increasing the oxygen saturation, please call Dr Raphael Beck or Dr. Amanda Barclay at the Pediatric Intensive Care Unit at the Victoria General Hospital at 727-4186.

VOMITING

If Shira is vomiting she may become dehydrated. This can be very serious for children with neuro-muscular weakness. Please contact Dr. Raphael Beck or Dr. Amanda Barclay at the Pediatric Intensive Care Unit at the Victoria General Hospital at 727-4186 or page Dr. Raphael Beck at 389-8925.

CONSTIPATION

If Shira has not had a bowl movement in 24 hours administer a vial of fleet baby lax rectally.

AEROSAL INHALERS DELIVERED THROUGH THE AEROCHAMBER



8AM
1 dose of Flovent

8PM
1 dose of Flovent

If you are not sure if the dose was given fully do not repeat. Wait until the next time you are to give Shira her neb.

COLDS

Ventolin X2 every 4 hours including 8am and 8pm

Protocol for Aerochamber use is on the next page.

FLOVENT HFA:

- ✓ NO CFCs
- ✓ SAME active ingredient
- ✓ SAME three strengths
- ✓ SAME dosing regimen
- ✓ SAME 30-day supply



FLOVENT HFA contains a medicine called fluticasone propionate, which is a synthetic corticosteroid. The natural corticosteroids in your body help fight inflammation. When inhaled regularly, corticosteroids also help to prevent symptoms of asthma.

VENTOLIN HFA can help

VENTOLIN HFA is an albuterol inhaler. Albuterol is a medicine that helps open the bronchial passages within your lungs to ease breathing during an asthma attack. **VENTOLIN HFA** can be used as a rescue medicine to help relieve your sudden asthma symptoms when they occur.

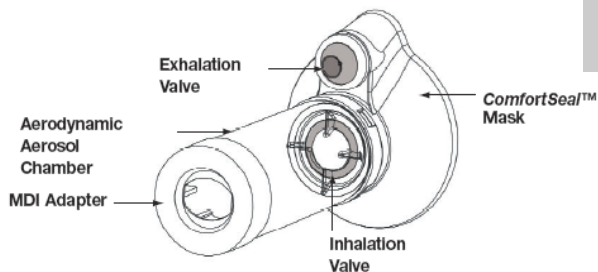
Like most asthma medicines and many other household products, **VENTOLIN HFA** uses a propellant — a gas that sprays the medicine out of the canister and into your mouth. But unlike some other asthma medicines, it does not use chlorofluorocarbons (CFCs) as the propellant. **VENTOLIN HFA** uses an alternative propellant called hydrofluoroalkane, or HFA. CFCs have been shown to deplete the ozone layer, and will be phased out for medical purposes as alternative propellants become widely available.

VENTOLIN HFA acts within minutes to help relieve bronchospasm and its symptoms for up to 6 hours. With **VENTOLIN HFA**, you can feel confident that you're using a medicine that's as effective as **VENTOLIN®** (albuterol, USP) Inhalation Aerosol and has a similar safety profile.

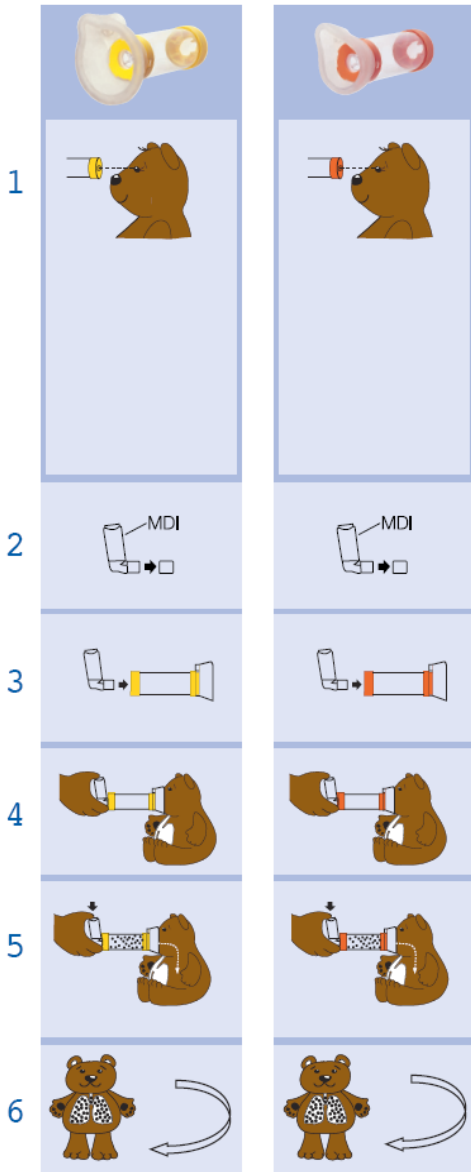
Ventolin HFA



Directions for Use (Single Patient Use Only)



AEORSPACER INSTRUCTIONS



ENGLISH

Carefully examine the product for damage, missing parts or foreign objects. Any foreign objects should be removed. The product should be replaced IMMEDIATELY if there are any damaged or missing parts. If necessary, use the inhaler (MDI) alone until a replacement is obtained. If the patient's symptoms worsen, please seek immediate medical attention.

Before use, make sure that instructions supplied with inhaler (MDI) have been read. Remove cap.

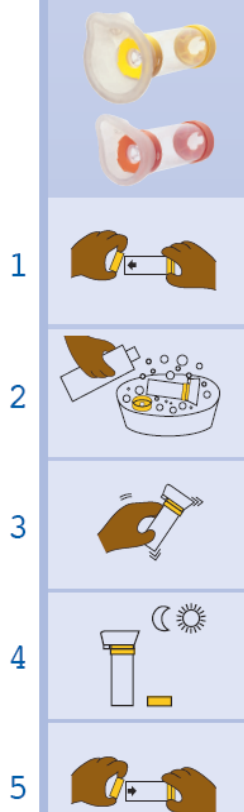
Shake the inhaler (MDI) well immediately before each use. Insert the inhaler (MDI) into the MDI adapter of the chamber.

Apply mask to face and ensure that there is a good seal.

Depress inhaler (MDI) at beginning of slow inhalation. Maintain seal for six (6) breaths after depressing inhaler (MDI). Administer one (1) puff at a time.

Follow instructions supplied with the inhaler (MDI) on amount of time to wait before repeating steps 3-5 as prescribed.

Cleaning Instructions



ENGLISH

Clean **AeroChamber Plus®** VHC only as per instructions before first use, then weekly.

Remove MDI adapter only. Do not remove mask and do not tamper with valve during cleaning.

Soak both parts for 15 minutes in lukewarm water with liquid detergent. Agitate gently.

Shake out excess water. **Do not rub dry.**

Let air dry in vertical position.

Replace MDI adapter when unit is completely dry and ready for use. Visually inspect per step 1 of the Directions for Use.

Cautions

- Ensure Directions for Use have been read prior to use and are kept available at all times.
- Do not leave **AeroChamber Plus®** VHC unattended with children. This is a medical device, not a toy.
- Do not disassemble the product beyond what is recommended in the Cleaning Instructions or damage may result.
- Review the use of this device with your healthcare professional prior to use.

Notes

- VHC = Valved Holding Chamber
- When not in use store in a suitable container.
- **This product contains no latex.**
- Product should be replaced after 24 months of continuous use. Environmental conditions, storage and frequency of use can affect product life span.
- If you have questions about the performance or usability of this product, please contact your healthcare professional.

Product Selection Guide

AeroChamber Plus® VHC	Infant Mask (0 - 18 months)
	Child Mask (12 months - 5 yrs.)
	Adult Mask or Mouthpiece (5 yrs. +)
AeroPEP Plus® VHC	Combination VHC and Positive Expiratory Pressure (PEP) product



Trudell Medical International™

Manufactured by: Trudell Medical International,
725 Third Street, London, Ontario, Canada N5V 5G4
E-mail: customerservice@trudellmed.com Web site: www.trudellmed.com

AeroChamber Plus®

Valved Holding Chamber with Infant/Child Mask



Replacement copies of this insert can be obtained by visiting www.trudellmed.com or by calling 1-866-510-0004

AeroChamber Plus® VHC Complies with CSA Standard Z264.1-02, Spacers and Holding Chambers for Use with Metered Dose Inhalers

THE  LUNG ASSOCIATION

The Lung Association believes that the use of products such as **AeroChamber Plus®** VHC with your metered dose inhaler is helpful in delivering medication to the lungs.

Patents Pending. Printed in Canada. P/N 101171-002 Rev.C September 2004
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CHEST PHYSIO THERAPY

UPPER ANTERIOR OR ANTERIOR SUPERIOR LOBES 100 CHEST PERCUSSIONS



MIDDLE LOBE LATERAL AND MEDIAL 100 PERCUSSION BOTH SIDES



TWO HANDED PERCUSSIONS COVERING FROM THE APICAL AND POSTERIOR TO THE POSTERIOR BASAL.



IMPORTANT

FOLLOW CHEST PHYSIO WITH COUGH ASSIST!!
During periods of Shira having a cold or adilectasys vibes should be incorporated into Shira's chest physio session focusing on the congested areas of her lungs. Use the stethoscope in the closet to evaluate Shira's lung capacity.

INDOOR STROLLER

QUICK Indoor

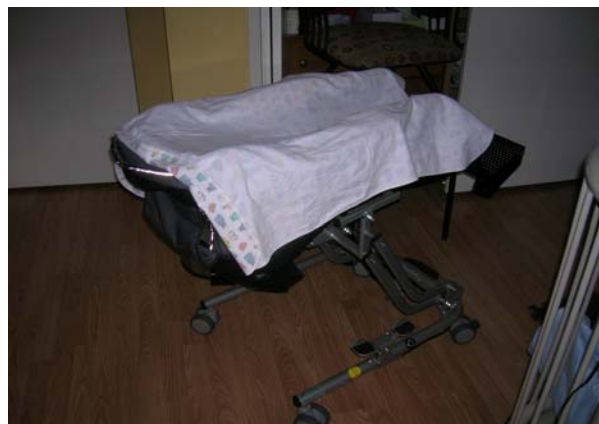


Exceptional positioning. Designed for safety. Built for **FUN!**

Use the **EASyS** seat from your LEX or MAX stroller with any **QUICK** hi/low base. Hi/low seating is a great way to promote social interaction with your child. Adjusting seat height and tilt-in-space are easy with the pneumatic shock -even with the child in the seat!



STROLLER SEAT FLAT

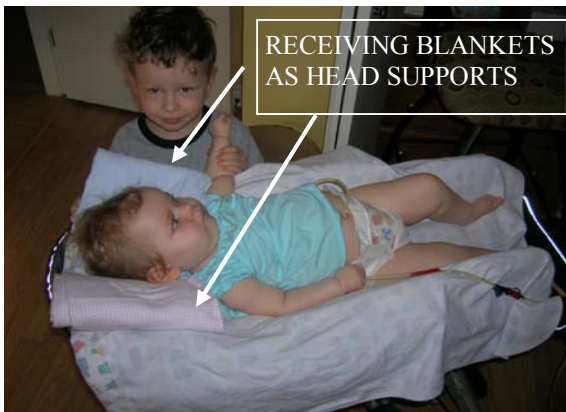


COVER SEAT WITH RECEIVING BLANKET



HEIGHT ADJUSTMENT UP AND DOWN

TILT—USED EXTENSIVELY TO HELP CONTROL SECRE-



OUTDOOR STROLLER



What makes JAZZ EASyS special is truly exceptional positioning possibilities.

Jazz EASyS is a special needs stroller that achieves many goals. The light-weight aluminum frame and seat are easy to fold and sport a stylish, non-medical look. JAZZ EASyS has the best comfort and safety features available, such as: cushiony ClimaBalance upholstery, [Balanced Safety System \(BSS®\)](#) stabilizing mechanism, dual spring seat suspension system, and reflective trim for nighttime visibility.

www.exomotion.com



ON BOARD EQUIPMENT FOR OUTINGS

- 1.) Oximeter
- 2.) Suction (extra suction catheters)
- 3.) Ambu Bag
- 4.) Feeding Pump
- 5.) DNR order (Do not resuscitate order)
- 6.) PICU Phone Number
- 7.) Diapers, wipes, blanket,
- 8.) Rain Screen



SLEEPING BAG

SUCTION MACHINE

OXYMETER

AMBU BAG
DIAPERS
WIPES
CHANGE OF CLOTHES

FEEDING PUMP

RAIN SCREEN