

# CONVENTIONAL AND NOT-SO CONVENTIONAL THERAPIES IN CF



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# OBJECTIVES

- ▶ To better understand the integrative approach to cystic fibrosis care- including complementary and alternative therapies
- ▶ To discuss how a multidisciplinary team can work with patients and families to improve adherence and even outcomes
- ▶ To discuss strategies for “integrating” non-conventional therapies to better manage both chronic and acute symptoms in cystic fibrosis.
- ▶ Become comfortable in discussing all therapies with the cystic fibrosis care team.



# WHAT IS CYSTIC FIBROSIS?

- Multisystem disease with a broad spectrum of severity
- Genetic disease with autosomal, recessive inheritance
- Progressive disease, especially chronic respiratory disease



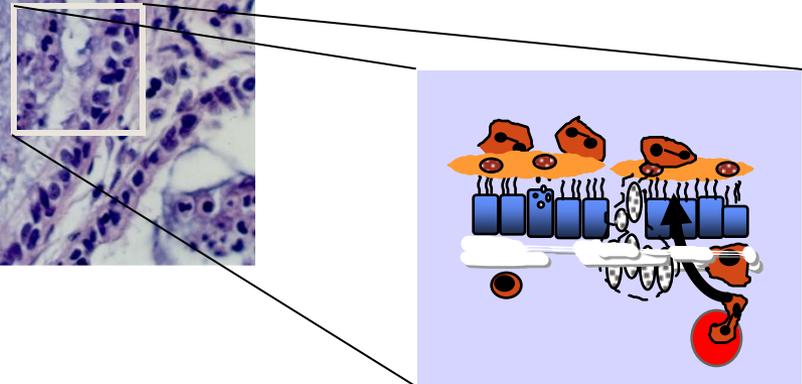
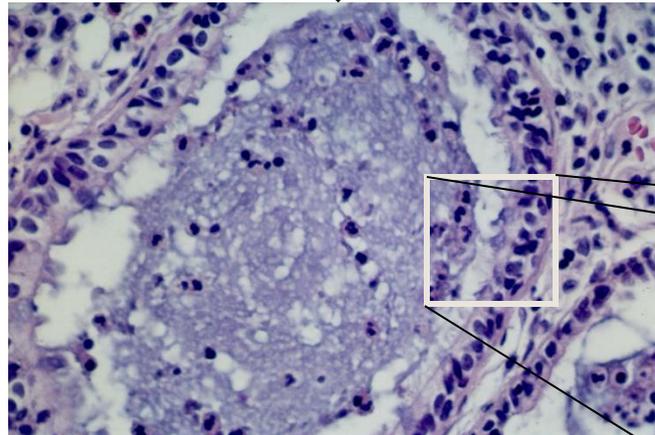
# PATHOPHYSIOLOGY OF CF

Modifier Genes

Abnormal CF Gene

Environment

- CFTR dysfunction or absence
- Infection with *P aeruginosa*
- Mucus plugging
- Inflammation



# CASE STUDY PRESENTATION

- ▶ Cheri is a 25 yr old who is delta F508, homozygous genotype and CFRD with liver involvement
- ▶ She works full time, graduated from college, married and requires ~ 1-2 hosp /year for a 7-14 day “tune up”.
- ▶ Currently has no increase pulmonary symptoms but feels “run down”. Would like “something” else to help maintain health, but not more “medications”.
- ▶ Presents to the clinic with questions about using complementary and alternative medicine
- ▶ Past history of good **adherence**.



# CONVENTIONAL THERAPY

- ▶ Duoneb nebulized 2/day
- ▶ Pulmozyme nebulizer daily.
- ▶ 7% hypertonic saline nebulized bid
- ▶ Vest bid
- ▶ Advair 250/50 2/day
- ▶ Rhinocort two sprays per nostril 2/day
- ▶ Sinus rinse daily
- ▶ Lantus, once a day
- ▶ Pancrecarb MT-8 capsules 2-3 per snacks, 6 with meals
- ▶ Ursodiol 300 mg 2/day
- ▶ Multivitamin daily
- ▶ ADEK daily.
- ▶ Vitamin K 5 mg daily.
- ▶ Azithromycin 500 mg 3/wk
- ▶ Tobi Podhaler 2/day every other month

Return to clinic every 3 months to have lung testing- is that enough?



# CASE CONTINUES

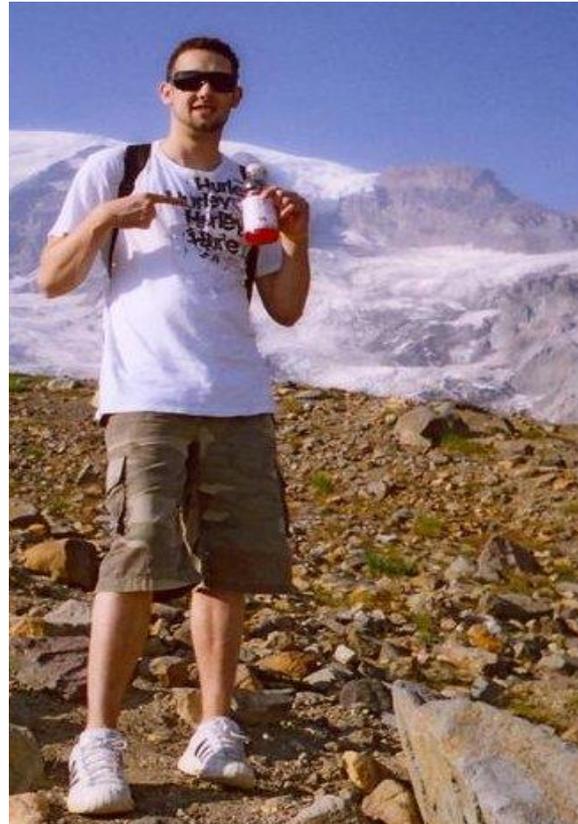


- Cheri is uncertain about the continual use of inhaled steroids, antibiotics and insulin
- She is interested in more “natural” approaches to CF therapy
- When asked which ones, she wasn’t sure but asked for the help of the CF team.
- What does the “team” say?
  - <http://www.drweil.com/drw/u/ART02973/Cystic-Fibrosis.html>
  - <http://www.greenmedinfo.com/blog/43-natural-alternatives-cystic-fibrosis1>
  - >1 million hits on Google!
    - About 1,070,000 results (0.39 seconds)





Remember-Cystic fibrosis can look like anyone-so therapies can also vary!



# CAM AND CF

- In a study of CAM use in 97 children and adults with CF (of which 81% of the respondents were parents of children, average age 11 yrs), 75% of patients used some form of CAM.
- The majority (53%) used biologically based therapies such as dietary supplements followed by mind-body medicine (49%) and manipulative therapies (14.5%).
- Although 77% of the patients who used CAM felt it was useful only 53% of these families reported use to their physicians.
- This study showed an increase use of CAM in patients with CF indicating the desire of parents and patients to have better control and self-management of their condition.



# ASSESSMENT

- ▶ Several factors are important to consider in determining your entire picture of health.
- ▶ Many chronic illnesses can be alleviated or improved after making certain changes in these areas.
- ▶ Nutritional, environmental and structural problems may seem physical in nature, these factors can greatly affect your mental and emotional health.
- ▶ Likewise, mental and emotional nurturing can positively affect the physical body.
- ▶ More than just what medicine to take or treatments to do....



# NON-CONVENTIONAL VS CONVENTIONAL

- Patients with chronic illness often reconcile the potential benefits of remedies and practices derived from different worldviews and understandings of human health and illness.
- CAM use in and of itself does not indicate irrational decision-making or unrealistic expectations of cure
- Consistent with taking responsibility for one's own health to the extent that it is possible in the context of a chronic disease.
- Consistent with an orientation toward doing what one can to improve one's own quality of life despite a disease.
- Resistance to non-conventional therapies could create a barrier to the optimal practice of self-care management.



# MAJOR CLASSES OF CAM

Enhance the mind's capacity to affect bodily function and symptoms

- ❖ Chiropractic
- ❖ Osteopathy
- ❖ Massage

Alternative medical systems

Mind-body interventions

Biologically-based therapies

Manipulative & body-based methods

Energy therapies

- ❖ Homeopathic
- ❖ Naturopathic
- ❖ Traditional Chinese and Ayurvedic treatments

- ❖ Dietary supp
- ❖ Herbs
- ❖ Foods
- ❖ Vitamins
- ❖ Natural substances

- ❖ Biofield therapy
- ❖ Bioelectromagnetic therapy



# TIME TO INTEGRATE: BUILD THE TEAM

- Start with a knowledge of each modality both by the practitioner and the family
- Discuss ideas and any therapies that are being used or considered.
- Just like there is with conventional therapies, there needs to be adherence and follow-up.
- Often just having the discussion of CAM helps improve adherence to conventional therapies
- Develop a plan, which can easily change

Textbook of Integrative Pediatrics (2009) Oxford Publishing



# WHERE TO START?

- Since 50-70% of patients with cystic fibrosis use CAM, the most common being herbal remedies/dietary supplements, need to address (omega 3 FA's #1 of all)
- For the patient who has mild to moderate disease, a step-wise approach in addition to conventional care might be considered.
- May include: Lifestyle, Nutrition, Supplements, Mind-Body therapies, Manipulative therapies and Alternative Health Care approaches (Traditional Chinese Medicine and Homeopathy)



# LIFESTYLE AND CYSTIC FIBROSIS

- CF is best treated early in an attempt to delay or decrease the progression of the disease.
- At times, is difficult, especially with CF since there are so many regular treatments daily
- Avoidance of allergens, tobacco smoke, and air irritants
- Sleep and good sleep hygiene can be important
- Exercise is a great activity-improves lung function, helps maintain weight and is an “anti-inflammatory” therapy.

YOUR  
Lifestyle



# EXERCISE AND CF- GOOD!

- ▶ Exercise used to be considered dangerous for CF patients
- ▶ Exercise is now considered a valuable tool for CF patients
- ▶ Natural form of chest physiotherapy-could use exercise as a substitute for “poundings” since the movements with exercise will help loosen mucus in the lungs.
- ▶ Exercise will often stimulate coughing, helping with the removal of secretions and better airway clearance
- ▶ Exercise also improves cardiovascular health and help with self esteem
- ▶ Requires guidance from the CF team.
- ▶ Ongoing study showing exercise can replace one “pound/day”.



# NUTRITION AND CYSTIC FIBROSIS

- ▶ Because diet is the major source of antioxidants, suboptimal intake during airway growth may lead to airway damage and reduced airway compliance.
- ▶ In chronic lung disease has been shown a beneficial association between fruits, vegetables, and other antioxidant-rich foods including fish.
- ▶ Along with a high calorie, high protein diet, patients with cystic fibrosis should increase their consumption of fruits and vegetables.
- ▶ Also, foods rich in omega-3 fatty acids while decreasing omega-6 fatty acid containing ones (vegetable oils) may decrease the inflammatory cascade in CF.



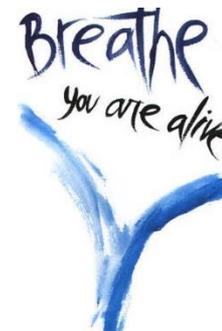
# MIND-BODY THERAPIES AND CF

- ▶ Mind-body therapies or cognitive behavioral therapies encompass several approaches and include relaxation therapy, breathing exercises, biofeedback, and hypnosis and guided imagery.
- ▶ The theory is that exercise helps decrease the inflammatory process that can be triggered by the autonomic nervous system through strong emotions.
- ▶ Stress has been associated with higher morbidity and cytokine levels attributed to airway inflammation.
- ▶ In addition to anxiety, stress is shown to influence the immune response and may promote increased sympathetic activity, and promote airway inflammation without overt symptoms



# MIND-BODY THERAPIES AND CF

- ▶ Self hypnosis, Breathing exercises including yoga, relaxation with or without guided imagery have all been studied in CF
- ▶ Recent study (Anbar) on imagery and sputum production without using any “devices”!
- ▶ Guided imagery and/or self-hypnosis has been shown to decrease shortness of breath or dyspnoea.
- ▶ May also help decrease chronic symptoms such as cough and decrease the use of certain medications such as albuterol
- ▶ Breathing exercises may also help with lung expansion especially breathing used in Yoga and martial arts





# SELF-HYPNOSIS AS A COMPLEMENTARY AIRWAY CLEARANCE TECHNIQUE IN PATIENTS WITH CYSTIC FIBROSIS

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F54

## Background

Common Airway Clearance Techniques for Patients with CF

- Manual chest physiotherapy (CPT)
- Active cycle of breathing techniques
- Autogenic drainage
- Positive expiratory pressure
- High-frequency chest compression (HFCC)
- Exercise

## A Working Definition of Hypnosis

Hypnosis is an altered state of consciousness characterized by a heightened state of suggestibility

## Hypnosis Does Not Involve:

- An unusual condition
- Sleep
- Placing the subject under control of the "hypnotist"

## Steps for Hypnosis in Clinical Practice

- Pre-hypnotic interview
- Induction
- Deepening
- Hypnotic Suggestions
- Trance Termination

**Effectiveness of Hypnosis for People with CF (n = 49)**

Number of Patients	Average Age (Years)	Achieved Goal (%)
32	17	67
50	17	50
100	20	100

## Methods

- Nine patients with CF
- Mean age 16 years, range 11-22 years
- All of the patients were using HFCC as their usual ACT
- The patients were instructed how to enter a state of self-hypnosis (SH) by using their imagination to experience a favorite place or activity, augmented by multisensory imagery
- The efficacy of the therapy was gauged by observation of any expectorated sputum and patient reports regarding their ongoing use of SH CPT

## Hypnotic Instruction

- Create a small imaginary character who can help clear your lungs by entering your airways and
  - (1) Remove sputum from one lobe at a time
  - (2) Spray hypertonic saline into one lobe at a time
  - (3) Photograph your airways in order to verify whether they are sufficiently clean. Then, clear any remaining sputum

## Results

- Among 7 regular sputum producers
  - 5 reported that SH CPT prompted expectoration of sputum > than produced during HFCC for 20 minutes
  - Time utilized for their SH CPT was 5-30 minutes
  - 5-75 mL of sputum was produced
  - 3 patients were observed to produce purulent sputum in response to SH imagined removal, and produced frothy sputum in response to imagined hypertonic saline
  - 4 patients produced purulent sputum only
  - 4 patients reported that they used SH CPT at home on a regular basis in addition to employment of HFCC

## Discussion

- Possible mechanisms of action
  - Induction of sputum production by encouraging imagined airway irritation that led to the development of cough
  - Imagery triggered autogenic drainage
  - Placebo response

## Conclusions

- The mechanism of action and long-term efficacy of SH CPT is unclear, and should be studied further
- As there is no down-side to use of SH CPT, this technique should be considered in patients with CF who produce sputum, as an adjunct tool to enhance the effect of traditional ACTs

## Reference

1. Anbar MD, Pediatr Pulm. 2006;30:463-4

## Additional Resources

- Anbar MD. Self-hypnosis for patients with cystic fibrosis. *Pediatr Pulmonol.* 2005;30:463-5
- Hume PA, Robinson EA, O'Sullivan BP, Fowler DJ, Wainwright NI, Wylie-Courand DA, White TB, Marshall AC. Clinical Practice Guidelines for Pulmonary Therapies Committee. Cystic fibrosis pulmonary guidelines: priority changes. *Chest.* 2006;129:1552-61
- Maysen DS, Anbar MD. Functional aspects of an upper respiratory disorder: cystic fibrosis. In: *Textbook of Hypnosis: Functional Hypnotic Disorders: When Hypnotic Symptoms Do Not Respond to Pulmonary Treatment.* New York, NY: Humana Press; 2012: 14-47.
- Ragan R, Mitzelmayr DA. Update in cystic fibrosis 2011. *Am J Respir Crit Care Med.* 2012;185:911-6



# DIETARY SUPPLEMENTS



- ▶ Herbal medications are classified as dietary supplements and are derived from plants.
- ▶ They be taken orally as pills, freeze dried capsules, or powders; used as tinctures or syrups; brewed in teas or decoction; or “applied” as salves, ointments, or poultices to the skin or mucous membranes.
- ▶ In survey studies, herbal supplements are often the most common CAM therapy reportedly used for the treatment of cystic fibrosis.
- ▶ A study of pediatric CF pts and families found 19% of patients were currently using supplements and 10% reported past use. 40% had not informed their physicians.



# DIETARY SUPPLEMENTS AND CF

- ▶ Common use has caused concern, because herbs may have interactions with conventional medications and even other herbal supplements.
- ▶ The FDA regulate dietary supplements as foods so they do not need to meet the same standards as pharmaceutical medications for proof of safety and effectiveness.
- ▶ Also, in many herbal supplements, the plant itself may not be characterized, active ingredients and standardization may not be known
- ▶ This makes the conventional scientific understanding of how herbal supplements might affect the body in conditions, such as asthma, difficult.
- ▶ Also reports of some supplements being contaminated with metals, unlabeled prescription drugs, microorganisms, and other substances



# WHAT'S IN THOSE SUPPLEMENTS

- The New York State attorney general's office **accused** four national retailers on Monday of selling dietary supplements that were fraudulent and in many cases contaminated with unlisted ingredients.
- Walmart, Walgreens, Target and GNC — which showed that roughly four out of five of the products contained none of the herbs listed on their labels.
- Walgreens: *Echinacea*: No Echinacea found but identified garlic, rice and daisy

The New York Times

February 3<sup>rd</sup>, 2015

<http://nyti.ms/1zATRgL>



# ENOUGH IS ENOUGH?

## Stupid Pills: The Politics of Fraudulent Dietary Supplements

NY Times, 2/6/2015

- Industry keeps growing, with 65,000 dietary supplements now on the market
- Consumed by nearly half of all Americans
- The larger issue is mistrust of authority, a willful ignorance that knows no political side
- Politicians promote a freewheeling market of quack products
- Others disdain modern medicine in favor of anything sold as “natural” or vaguely countercultural



# STUDIED SUPPLEMENTS

- ▶ In cystic fibrosis, asthma and other chronic lung diseases, antioxidants such as Vitamin C, selenium, and fish oil/omega 3 fatty acids have been studied with variable results
- ▶ Taurine, an essential amino acid may improve fat absorption and weight gain in CF (studies-80's)
- ▶ N-acetylcysteine (NAC) may soon be “conventional” care for the anti-oxidant properties it provides.
- ▶ Creatine has been studied in CF and shown to improve muscle strength, at least short term.
- ▶ There are so many one might think about taking.....



# DIETARY SUPPLEMENTS FOR CYSTIC FIBROSIS

- ▶ **Probiotics**
- ▶ B complex
- ▶ Extra Vitamin C, E, A
- ▶ Calcium, Vitamin D
- ▶ Magnesium
- ▶ Ginger, ginko, **garlic**
- ▶ Echinacea (not routinely)
- ▶ **Essential oils for inhalation (eucalyptus, oregano)**
- ▶ Lipoic acid (esp for CFRD)
- ▶ Milk thistle
- ▶ **Tumeric**
- ▶ **Grapefruit seed extract**
- ▶ Baker's yeast, Green tea
- ▶ Airborne: 18 ingredients
- ▶ Coenzyme Q-10
- ▶ Blue green algae
- ▶ Spirulina, alfalfa
- ▶ Pine bark (Pycnogenol)
- ▶ Gingseng
- ▶ Licorice
- ▶ **Boswellia**, bromelain
- ▶ Cayenne, mullein, horehound





# PROBIOTICS STUDY IN CF

- Intestinal inflammation is a typical finding in CF patients and bacterial overgrowth may be present and studies suggest a constant intestinal inflammatory state
- Probiotics are live bacteria administered orally, that may decrease severity of GE, and prevent eczema in children
- The probiotics may help improve intestinal barrier function and modify immune response.
- 10 CF patients with mild–moderate lung disease and PA colonization, were treated with probiotics for 6 months (Lactobacillus GG): PFT's did not change post-treatment. No change in sputum bacteria, neutrophil count, and IL-8 levels was observed.
- Did find a reduction in pulmonary exacerbations rate



# MANIPULATIVE THERAPIES IN CF

- ▶ Manipulative therapies include osteopathic manipulation, chiropractic manipulation and massage
- ▶ Children with CF and their parents have reported a positive response to massage therapy with reduction in anxiety, improved mood and increase in peak flow measurements have been shown in small studies.
- ▶ Therapeutic massage can help drain mucus from the lungs
- ▶ Osteopathic manipulation- 5 main categories with over 100 different individual maneuvers



# TRADITIONAL CHINESE MEDICINE (TCM)

- ▶ TCM uses the belief in an unseen vital energy that affects patients' health and how this energy, or qi (chi), flows through the appropriate channels is monitored
- ▶ Practitioners can affect this flow or intensity by manipulating its balance using acupuncture, Asian herbs, diet, and physical therapy.
- ▶ Cystic fibrosis has not been recognized in Asian medicine but there are traditional herbal formulas used for all of the symptoms and possible deficiencies or excesses seen in CF



# ACUPUNCTURE?



- ▶ Another modality often used is acupuncture for chronic lung problems including CF
- ▶ However, most studies have not shown improvement in such diseases as asthma or COPD.
- ▶ Some practitioners feel that acupuncture alone may not be beneficial since it is just one part of TCM, and TCM is a holistic approach to a chronic problem
- ▶ One study at Boston's Children's showed acupuncture to be effective in decreasing pain complaints in patients with cystic fibrosis



# HOMEOPATHIC MEDICINE

- ▶ **Homeopathic medicine** is a belief that "like cures like"
- ▶ This **Law of Similars** means that small, highly diluted quantities of medicinal substances are given to cure symptoms, when the same substances given at higher or more concentrated doses would actually cause those symptoms.
- ▶ The most dilute remedies are the strongest (**Law of Dilution**)
- ▶ Before prescribing a remedy, homeopaths take into account a person's constitutional type: physical, emotional, and psychological makeup (**classical vs clinical**)
- ▶ The remedies can be mineral, plant or animal



# HOMEOPATHIC NOTATIONS



- ▶ The notation 6X means that the active substance is diluted 1:10 in a water-alcohol mixture and succussed.
  - ▶ This procedure (diluting and succussing) is repeated sequentially six times.
  - ▶ The concentration of the active substance is one part in ten raised to the sixth power, or one part per million.
- ▶ The notation 30X means the 1:10 dilution, followed by succession thirty times.
- ▶ Homeopathic physicians agree that not a single molecule of the active substance could remain in many of their remedies. The process of succussion is presumed to charge the entire volume of the liquid with the same memory.



# PUTTING IT ALL TOGETHER



- ▶ Conventional therapies including airway clearance, nutritional support and anti-inflammatories
- ▶ May use some supplements in place of conventional meds after discussing with your health care team (bromelain, pine bark, grapefruit seed extract, garlic) along with anti-oxidants (Vitamins, selenium, NAC) and alpha lipoic acid (CFRD)
- ▶ Consider yoga, martial arts (Tai Chi), meditation
- ▶ Exercise!
- ▶ If interested, then TCM, homeopathy or naturopathic medicine but need to be able to communicate with all the providers so care is “transparent”.



# INTEGRATIVE MEDICAL PLAN FOR CF

- Medications- what are the most important and how to improve adherence, discuss concerns about side effects and cost
- Lifestyle changes including exercise
- Nutrition and dietary changes
- Supplements including herbal remedies
- Possible use of TCM, Manipulative therapies and homeopathy
- Most important: education and teamwork in regard to all aspects of the health care plan
- Remember: Mindfulness includes attention, intention and attitude



# THAT'S ENOUGH-THANKS!





<http://newdaywell.org/home.html>



<http://www.cflf.org/>



<http://runsickboyrun.blogspot.com/>

<http://www.cfww.org/about/>



# A GUIDE FOR USING CAM

## Government sites

- ▶ National Center for CAM:

<http://nccam.nih.gov/>

- ▶ Office of Dietary Supplements:

[http://ods.od.nih.gov/Research/CARDS\\_Database.aspx](http://ods.od.nih.gov/Research/CARDS_Database.aspx)

- ▶ MedWatch:

<http://www.fda.gov/medwatch>

- ▶ CAM on PubMed:

<http://www.nih.gov/news/pr/feb2001/nccam-05a.html>



# A GUIDE FOR USING CAM

## Academic sites

- ▶ Boston-Longwood Herbal Task Force:

<http://www.longwoodherbal.org/>

- ▶ Ohio State University

- ▶ <http://wexnermedical.osu.edu/patient-care/healthcare-services/integrative-complementary-medicine>

- ▶ Beth Israel Medical Center NYC:

<http://www.healthandhealingny.org/>

- ▶ University of California, Berkeley Newsletter:

<http://www.wellnessletter.com/html/ds/dsSupplements.php>

Sloan Kettering: <http://www.mskcc.org/cancer-care/integrative-medicine/about-herbs-botanicals-other-products>



# COURSES AND WEBSITES

- ▶ Herbs and Dietary Supplements-Wake Forest
  - ▶ <http://www.wfubmc.edu/Research/Center-for-Integrative-Medicine-Research.htm>
- ▶ Botanicals and Health series-University of Arizona
  - ▶ <http://integrativemedicine.arizona.edu/education/index.html>
- ▶ Nutrition and Health Conference (12<sup>th</sup> annual)
  - ▶ <http://nutritionandhealthconf.org/>
- ▶ Consumer Labs
  - ▶ <http://www.consumerlab.com>
- ▶ Natural Medicines Comprehensive Database
  - ▶ [http://www.therapeuticresearch.net/\(ctt54155qny2vs55ryiv2d32\)/home.aspx?li=0&st=0&cs=&s=ND](http://www.therapeuticresearch.net/(ctt54155qny2vs55ryiv2d32)/home.aspx?li=0&st=0&cs=&s=ND)
- ▶ Quackwatch
  - ▶ <http://www.quackwatch.com>



# THANKS!

