KAPOSI SARCOMA CLINIC
UC Department of Dermatology

# OUTLINE OF STAGING PROTOCOL FOR K.S. PATIENTS

## I. PATHOLOGY

A copy of the diagnostic path slide is sent from the referring physician to Dr. Richard Sagebiel (666-1543) for a second reading.

## II. INTAKE INTERVIEW

With Helen Schietinger, RN, Clinic Coordinator, for admission and orientation to the clinic and initiation of staging workup. (666-1407).

## III. CLINIC APPOINTMENT

For history, physical and complete dermatological exam with Dr. Marcus Conant, Co-Director of K.S. Clinic.

## IV. STAGING WORKUP

1. Abdominal CT Scan
2. Upper and lower endoscopies
3. Opthamological exam for cotton-wool spots and retinal abnormalities
4. Oral exam by Oral Medicine for oral and pharyngeal lesions
5. Pulmonary function tests to screen for possible pulmonary abnormalities indicating pneumocystis
6. Chest X-ray
7. Lab tests
   a. Stools for ova and parasites
   b. Stool culture
   c. Rectal and throat swabs for G.C.
8. Possibly lymph node biopsy, if lymph nodes are swollen
9. Possibly further skin biopsy
10. Blood work
    -- CBC, Sed rate, SMA-23, RPR, ANA, Platelet count, Rhuematoid factor, Heterophile
    -- Hep BS Ag (if positive, order HBe Antigen)
    -- Hep B Core Ab
    -- Hep BS Ab
    -- T Helper/Suppressor ratio
    -- HLA typing (A, B, C and DR)
    -- Viral cultures (semen and urine) and antibody titers for CMV
    -- EBV antibody titers

WHAT IS KAPOSI'S SARCOMA?

Kaposi's sarcoma is a cancer of tiny blood vessels, named for the medical researcher who first isolated the disease. The vessels usually affected are in the skin, but the disease may also infect the lymph nodes, organs of the body, or the mucous membrane of the GI tract.

WHAT WILL THIS CLINIC DO FOR AND TO YOU?

Helen Schietinger, RN, is the Nurse Coordinator of the Clinic. Her phone number is 666-1407. Dr. Marcus Conant is the dermatologist, Dr. Paul Volberding is the oncologist, and they are Co-directors of the Clinic. Nancy Beckman is another nurse working with the Clinic, Dr. Steve Mehalko is a doctor who works with us, and Paul Dague, PhD, is the psychologist.

Over the next two weeks, you will be given tests to identify the extent of your disease and to determine what course of treatment is best suited to your case. You will also take blood tests which will measure the "health", or degree of effectiveness, of your body's immune system. We will also examine your blood samples to determine if you have been exposed to certain viruses, as well as measuring various other characteristics of your blood.

When you have completed all the tests, the doctors will determine which treatment they recommend as best for you, and you will probably then begin a program of weekly chemotherapy treatments.

There are two types of chemotherapy given to Kaposi patients. The first, a one-drug treatment, is given patients whose cases are deemed "slowly progressive." The second, a three-drug treatment is given those patients whose cases are judged to be "rapidly progressive." The oncologist, or cancer doctor, Dr. Volberding, will tell you specifically about these treatments, the drugs and their known side effects. Please do not hesitate to ask <u>any</u> questions you may have.

As you go through the initial testing and the subsequent treatment, you will be asked to participate in a study we are conducting to learn more about the possible cause of Kaposi's. Medical science knows relatively little about the causes of Kaposi's at this time. Participation in this study is entirely voluntary, and you may take the prescribed treatments without participating. Your participation in the study will permit us to collect any necessary records from your medical records on file elsewhere, to question you about your medical history, and to compare your case with those of other patients.

Whether or not you participate in the study, you will be given regular blood tests, as well as necessary laboratory and x-ray exams throughout your course of treatment to monitor your condition and response to the treatment.

Before you participate in the study, you will be given a consent form to read and sign.

KAPOSI'S SARCOMA AND IMMUNODEFICIENCY--WHAT IS THE CONNECTION?

One reason the study we are conducting is so important is the need to learn what the connection is between the occurrence of Kaposi's and defective immune systems in the patients. Kaposi's is not a new disease, having been found among older Italian and Jewish men in particular, for well over a century and among certain Africans. But along with some other diseases such as Pneumocystis carinii pneumonia, yeast and fungal infections, it has now been detected recently in a significant number of gay men, and also in both heterosexual men and women of no particular ethnic background, and with no apparent previous disease that would leave them susceptible to these diseases.

Virtually all the recent patients suffering from these diseases have been found to have impaired or defective immune functions. That is, the cells in the blood and lymph systems which help to fight infection at the cellular level are not present in the right numbers and the proper proportions to enable these patients to fight off Kaposi's and these other infections, which usually appear in persons who have been severely or chronically ill. The evidence indicates that you may have Kaposi's because your body's ability to protect itself from foreign substances and infections has somehow been decreased.

WHAT CAN I DO ABOUT MY IMPAIRED IMMUNITY?

Because your immune system is not working properly, you need to take extra precautions to protect yourself:

1. Get plenty of sleep

2. Seek ways to manage and control stress in your life, and to cope with the stress which comes your way. The Shanti Project is an excellent resource for learning new ways to handle stress. We will give you the Shanti telephone number.

3. Eat nutritious food, including fresh fruits and vegetables every day. If you are not sure whether or not your diet is healthy, speak to the nurse about it.

4. Because evidence suggests that most recreational drugs affect the immune system, take a close look at your drug consumption. Both alcohol and marijuana consumption may aggravate suppressed immunity, but other drugs may also create problems or increase existing problems. Smoking tobacco may be more harmful for an immunosuppressed person than for others.

During this time, you may come under more emotional stress than you are accustomed to coping with, and you might ordinarily turn to drugs, including alcohol. If you have difficulty cutting down or stopping, don't be embarrassed or frightened. Ask the nurse or psychologist about getting help and support.

5. You are more susceptible to any infection when your immunity is impaired. If you are having sex with more than one person, and these partners are not known to you, you are at great risk of getting any number of sexually transmitted diseases at a time when your body is ill-prepared to deal with them. Because of this, and because of the possibility of transmissibility which will be discussed in the next section, it is suggested that you not be sexually active with more than one person during this time of testing and treatment. We are saying this only because of the risk of infection, not because there is anything inherently harmful in any sexual practices. Sexual activity itself, heterosexual or homosexual, does not cause disease.

The research done to date has pointed up some "risk factors", or things that many of the patients have in common that are not usually found in non-patients. For example, many of the patients have been frequent or heavy users of recreational drugs. Many have also revealed a history of active sexual activity with many different partners. A number of patients have been men who have participated in fisting. None of these activities is characteristic of all the patients, and none can be said to be a cause of Kaposi's or pneumocystis pneumonia, but any could contribute, indirectly, to depressing the immunity.

IS KAPOSI'S CONTAGIOUS?

Kaposi's sarcoma in and of itself is not contagious. However, whatever is impairing the immune system may be transmittible, and this is a serious concern.

One theory about why these diseases are appearing now with greater frequency is that the incidence of sexually transmitted diseases has risen in the past few years. The reported incidence of all types of hepatitis, viral infections such as cytomegalovirus (CMV), Epstein-Barr Virus (EBV), and herpes, intestinal parasites (amoebas, giardiasis, etc.), as well as the classic venereal diseases, gonorrhea and syphilis, has risen in recent years. The theory maintains that a sexually active gay man may expose himself to a great many more infections than he would have a few years ago, and that at some point his immune system may simply "give up" or become overloaded.

Another theory maintains that there may be a new strain of virus or other organism that depletes the immune system, and is itself sexually transmissible. One such suspect is the previously mentioned CMV, which is very prevalent in the gay male community.

The upshot can be simply stated: we do not know for sure if Kaposi's is contracted because a transmissible infectious agent has depleted the immune system. However, this is a possibility, and consequently we suggest that you not have sex right now. If you do have sex during this period, it is best for you to stick with one partner known to you, known to be disease free, and fully informed about your condition. It would not be fair to a lover, friend, or any sexual partner to expose him to the possibility of becoming immunodeficient when he is not informed. Also, you should avoid sexual activity in which your semen is swallowed by your partner or received into his rectum (you can use condoms to avoid this), and in which you are the passive partner in anal/oral sex. This will hopefully protect your partner(s) from transmission of any agent that may be responsible for depleting the immunity.

Rough draft

UCSF Department of Dermatology
Kaposis Sarcoma Clinic-Mondays, 11 AM-1 PM
Derm Clinic
New Patient Protocol

I. Clinical history- The name, address and phone number of the referring physician(s) is most important. A letter is to be dictated to this physician as the conclusion of the initial visit, and a phone call on the same afternoon, if possible. This will go far towards developing rapport with and continued referrals from the community. Also, a determination should be made as to whether the referring M.D. wants us to act as a treatment center for the patient, or to merely suggest therapy.

The usual clinical history ( history of present illness, past medical history, review of systems, family history and social history) is to be obtained. Any indepth Epidemiological questioning should be deferred, as the CDC on their visits will be responsible for this aspect of the history.

II. Physical Exam- A complete physical is to be done on the initial visit, as well as a body stamp with representation of all lesions, with size, duration and dermatological description. Also, please photograph the patient's lesions.

III. Lab studies- These are most important, and every effort should be made to see that all of our patients have these studies completed as our baseline data-further tests to be done during therapy (as follow-up visits) will be distributed later. If possible, all of the following tests should be completed before commencing therapy.

Routine studies:
- CBC
- Sed rate
- SMA-23
- YDRL
- ANA
- Stool guiac
- Stool (X3) for ova and parasites
- Throat culture for GC
- Rectal culture for GC
- HepBS Ag
- HepBS Ab
- HepBcore antigen
- HepBcore antibody
- ? Latex fixation or fixative ?

IV. Radiologic studies-
- Chest X Ray
- Abdominal CAT scan
- GI series with barium enema
- Nuclear medicine body scan (cesium-Tc)

V. Consults- Opthalmology-to rule out CMV retinitis-these should be referred to Dr. Friedlander

ENT-for indirect laryngoscopy to rule out naso-

pharyngeal lesions
GI-for endoscopy, protoscopy and colonoscopy-these should be referred to Dr. Dave Altman

VI. Special studies- 1. Biopsy*-all larger lesions which have an excisional biopsy should be divided, gently, into S parts-for routine histopathology (in formalin) plastic embedding (in paraformaldehyde), EM (in glutaraldehyde), and the fourth part frozen in $LN_2$ (wrapped in tin foil and placed in labeled tube), and the fifth part (1-2$mm^3$) placed by sterile technique into a sterile container with a small amount of sterile saline-to be transported within the hour to Dr. Drew for viral culture (Mt. Zion). Punch biopsies should be divided, gently, between routine histopathology and EM. *Please make an effort to contact Drs. Van Fletcher, Scott McNutt or Richard Sagebiel before a patient is to be biopsied-for best coordination.

2. HLA typing-This study is done in the Immunogenetics Lab of Dr. Garavois (ext. 3883, 520 HSE)
Please notify him in advance of any patient's specimens that will be sent. He will freeze the cells to be seen at a later date, and needs 20-30 cc of blood in yellow-topped tubes (which contain ACD). These can be obtained from the kidney transplant unit in Moffitt (call ahead).

3. Viral titers and cultures-These are being seen at Mount Zion Hospital, by Dr. Larry Drew (567-6600 ext. 2000). Dr. Drew needs 1-2 red top tubes for this study. In addition he will be running cultures on the biopsy material (see section VI:1), and possibly on other body fluids.

4. Immunology lab (UCSF) tests- 5th floor HSE, ext. 1712
Dr. Konrad Kasavant
T & B cell markers- please call ahead and notify lab
35 cc of blood in lavender-topped (EDTA) tubes
CH50, C3, C4, and Immune Complex Screen- 2 red top tubes (these must be delivered to the lab within one hour or the test in invalid)
Quantitative immunoglobulins-one red top tube (request IgM,IgG,IgA,IgE)

Other studies-Lymph node biopsies should be processed for routine H & E, EM and plastic embedding, and 2 small portions frozen-one for future studies, one for possible estrogen receptors.

5. T subset determination- by Dr. Stobo's lab ext. 1291 At least initially we should call this lab, when patients arrive, lab will send over a technologist to draw the blood.
6. PHA stimulation and mixed lymphocyte culture- Dr. Diane Wara's lab ext. 4860 (Nanita)- These are drawn and run only on Tuesdays, so schedule with Casey on ext. 2171 on Monday.

VII. Skin tests- Trichophyton
PPD
Carsdida
Mumps
(SK-SD)

Conclusion: Although the lists of tests is, at first appearance, formidable, there will soon be a nurse hired to coordinate.