

## CANCER AND THE RISK OF X-RAY EXPOSURE

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Cancer poses a major risk for A-T patients. It may also be a small risk for carriers of A-T (see Inheritance, Chapter 3). These two issues will be discussed separately.

### Cancer in A-T Patients

**Cancer risk:** Studies over the past 30 years suggest that somewhere between 10 to 30 percent of all A-T patients will develop a cancer sometime in their lives. The vast majority of these are cancers of the white blood cells called lymphocytes (the cancers are called lymphoma and leukemia). These cancers can occur at any age in A-T patients, though they occur in greater numbers when patients are over the age of 10 years. It is difficult to provide a simple list of the presenting symptoms of these cancers. In general, lymphomas first appear as masses or swellings in areas of the body where there are lymph nodes (neck, chest and abdomen) or cause persistent fevers without other explanation. Leukemias typically cause bleeding, bruising, fevers, pale appearance, malaise and bony pain on occasion.

**Screening:** For leukemias and lymphomas, the treatment and prognosis are not very dependent on when the tumor is diagnosed. In contrast to cancers of the breast, colon, lung, etc., there is not a critical need to diagnose these types of cancer at the earliest possible time. Thus, screening tests are not very useful. At most, it is reasonable to obtain a routine Complete Blood Count (CBC) at times when other blood samples are being taken, or perhaps on a yearly basis. More important is that the pediatrician or primary doctor be aware of the propensity of A-T patients to develop these tumors, and to consider them in the differential diagnosis when symptoms arise.

**Treatment:** Leukemias and Lymphomas are *TREATABLE* and effective chemotherapy agents are available that can be utilized in A-T patients. A discussion of the agents and doses to use must be individualized based upon the specific tumor and the patient's general health. It is strongly recommended that the treatment of cancer in an A-T patient be supervised by a highly qualified specialist who works at a major cancer treatment center.

**Prevention issues:** Prevention of cancer is always the best treatment strategy. This is possible for some cancers when risk factors are related to behavior (for example, preventing exposure to cigarette smoke will reduce the risk for developing

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lung cancer). Unfortunately, the leukemias and lymphomas that arise in A-T patients probably arise spontaneously from lymphocytes that break their DNA naturally, and not because of exposure to anything in the environment. We actually do not know the real risk of radiation in A-T patients in terms of cancer risk, but it would be expected that natural or medical exposures to radiation carry only a very small risk.

It is not likely that even incredibly high doses of vitamins or anti-oxidants would affect the occurrence of cancers in the lymphocytes of A-T patients. In fact, mega-doses of some anti-oxidants could potentially be harmful in A-T patients. We recommend a well-balanced diet supplemented as necessary with a daily multivitamin.

### **Risk of X-Ray Exposure**

The amount of radiation from medical x-rays is quite low and the actual cancer risk from medical x-rays to A-T patients and carriers is unknown but likely to be extremely low. However, it makes sense only to do medical x-rays when it is absolutely necessary. In other words, x-rays should be done only when the result is going to affect a medical treatment decision. For example, if a child breaks his arm, an x-ray will be necessary to adequately treat him and the benefit clearly outweighs the risk. However, one should not use medical x-rays for screening purposes. As an example, routine screening dental x-rays would not be desirable, but a dental x-ray to evaluate tooth pain would be reasonable.

#### **GUIDELINES FOR USE OF DIAGNOSTIC X-RAYS IN A-T**

- Avoid x-rays when possible
  - Routine screening dental x-rays should be avoided
  - Sometimes another test (MRI scan or sonogram) can provide equivalent information without x-rays
- X-rays are reasonable when the result will affect medical management
  - If you/your child have fever and cough, and the doctor hears sounds characteristic of pneumonia, antibiotics can be prescribed without need for a chest x-ray
  - If symptoms persist despite antibiotics, a follow-up chest x-ray may be useful

Radon is a natural form of ionizing radiation that comes from the ground and tends to be at high levels in some geographic areas of the country (and usually just in basements with poor ventilation). It makes sense to have a house tested for radon and, if your house tests high, a pump can be installed to alleviate the problem. There is no data to suggest that A-T patients have a higher risk for radon-induced cancers than other individuals.

A-T patients are sensitive to sunlight (UV radiation), but their sensitivity is not really in terms of a high incidence of skin cancer. Because of thin skin and premature skin aging, A-T patients tend to have very dry skin that burns very easily in the sun. Thus, it is wise to use high-grade sun block and hats and sunglasses in intense sun, but do not use this sensitivity as a reason to keep your child indoors! A-T patients can enjoy the outdoors as much as anyone and should be encouraged to do everything they are capable of doing, indoors and outdoors.

## Cancer in A-T Carriers

**Cancer Risk:** Epidemiologic studies have uncovered an increased risk of breast cancer in women from families in which there is an A-T patient. Studies are continuing to try to evaluate the actual increased risk of breast cancer in the general population in women who are A-T carriers. Unless evidence is presented to the contrary, it is probably wise to assume, for now, that there is a small increased risk of breast cancer in A-T carriers and act accordingly. Interestingly, the statistically significant increased breast cancer risk in A-T families appeared to occur in women over the age of 60. This should be kept in mind for screening recommendations. If new data arises that suggests increased cancer risks in younger individuals who are A-T carriers, the screening recommendations may change.

**Screening/early diagnosis:** Early diagnosis of breast cancer can have a significant impact on treatment and outcome. Thus, it is desirable to have an effective screening program to provide early diagnosis in individuals at high risk for breast cancer. However, the possibility that the increased risk in A-T carriers occurs when they are over the age of 60 years alters this recommendation. The current common test used for screening for breast cancer is a mammogram. This is an effective test, but it does require a very small exposure to ionizing radiation, and it is conceivable that ionizing radiation could increase the risk of breast cancer in A-T carriers. Thus, the question arises whether this is the most effective screening test for A-T carriers. The common practice in this country for women in the general population is to recommend an initial screening mammogram between the ages of 35 and 40, mammograms about every 2-3 years at ages 40-50, and then yearly mammograms after age 50. Since the greatest risk of breast cancer in A-T carriers may be over the age of 60, it may not make sense to increase the frequency of screening mammograms at younger ages, as is recommended for persons with other breast cancer susceptibility genes. At this time, the routine schedule of mammographic screening is recommended for A-T carrier women. This recommendation could change if new epidemiologic data demonstrates a significant increased risk at younger ages.

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One screening test that all women who are A-T carriers should do regularly at all ages is a self-screening examination. This is the most effective screening method since it can safely be done each month and any worrisome findings can easily be followed up by other tests. Every female A-T carrier should do *MONTHLY* breast self-examinations. If you are not expert at performing them, have your doctor teach you how to do them well.

### **Risk of X-ray Exposure**

It is well documented that ionizing radiation increases the risk of breast cancer, but it is unclear how much exposure to medical x-rays would increase the risk of breast cancer development in A-T carriers. Nevertheless, as discussed for A-T patients, it makes sense to limit medical x-ray exposures to those situations which are absolutely necessary. In other words, diagnostic x-rays should be performed only when medical treatment decisions could be altered by the test result. It is not necessary to avoid them at all costs, but it makes sense to reduce the number of x-rays if possible, especially those in which the breast tissue would be in the radiation field, such as a chest x-ray. As with A-T patients, there is no obvious benefit to be gained from mega-doses of vitamins, but well-balanced, low-fat diets and one-a-day vitamins are reasonable. And whatever you do, **DON'T SMOKE!**

### **Recommendations for Identifying A-T Carriers**

The question frequently arises as to whether family members, such as aunts, uncles, cousins and siblings should be tested to determine whether they are carriers. This is a very complex question and it would require an extensive personal discussion to elaborate on all of the individual issues that should be considered. Since we do not have effective prevention or intervention strategies for the potential breast cancer risk, however, it is not clear how much benefit arises from knowing that someone is an A-T carrier. This is especially true since the breast cancer screening recommendations at this time are not really different for A-T carriers compared to the general population (see screening recommendations above).

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