

Head & neck squamous cell carcinomas in FA.

I have heard that if you have Fanconi Anaemia, you are at increased risk of ‘head & neck cancers’...is this true?

Fanconi Anaemia affected individuals are at increased risk of developing cancer of the mouth and throat (oral cavity/oropharyngeal cancer) and also, more rarely, around the genital/back passage area (anogenital cancer). The specific cancer type is known as squamous cell carcinoma. These cancers are usually an issue for FA-affected adults but have been reported in the teenage years. Such cancers are more difficult to treat with chemotherapy and radiotherapy than similar cancers in non-FA affected individuals. Treatment is therefore usually surgical removal. Thus it is important to identify any cancers early, perhaps even as ‘pre-cancers’, i.e., before any cancerous cells have had a chance to invade into the underlying soft tissues (also known as ‘in situ’ cancer). The earlier any cancer or pre-cancer is identified, the easier it is to surgically remove.

What can I do as an FA-affected individual to reduce my risk of developing oral cavity/oropharyngeal cancer?

Good dental hygiene, regular check-ups with the dentist, and avoiding smoking and consumption of alcohol are all known to reduce the risk of oral cavity/oropharyngeal cancer. In addition, infection with a virus known as Human Papilloma Virus may also contribute to the development of such cancers. Thus the following recommendations can be made:

- ***HPV vaccination should be done on all FA-affected individuals from the age of one year or as soon as the diagnosis of FA has been established. HPV vaccination should be repeated at an appropriate time after any bone marrow transplant. Note that HPV vaccination is not a substitute for an appropriate cancer screening programme in the FA affected individual.***
- *FA affected individuals should avoid alcohol consumption (including in some oral mouth washes) and smoking/tobacco products. Parents of FA individuals should not to expose their children to passive smoking.*

- *FA affected individuals should have six monthly dental check-ups with an NHS dentist or equivalent and should maintain good oral hygiene (tooth-brushing twice daily, dental flossing, use of non-alcohol based mouthwashes).*

What about screening for such cancers?

Early detection of any cancerous area is important, thus regular checks by a specialist are required. Self-examination is also important. If you notice any white patch, or any other area of abnormal appearance, texture, or consistency in the mouth, or any new neck swelling, get it checked out as soon as possible. Ensure that your doctor knows that any referral made for a worrisome problem is a 'two week wait' (a 'two week wait' referral is for suspected cancer...the specialist to whom you are referred to has to see you within two weeks of receiving the referral, even if it doesn't turn out to be cancer). In particular, the following is recommended:

- ***All FA affected individuals from the age of 10 years should have four times a year screening for oropharyngeal cancer. Screening is not necessary below this age.***
- *Screening for oropharyngeal cancer in FA should be done by a head & neck cancer specialist who is a core member of a regional head & neck cancer MDT, who has demonstratable experience in head & neck cancer screening, and who has at hand all available techniques concerning cancer screening including endoscopy (pharyngolaryngoscopy).*
- *A child with FA undergoing screening for oropharyngeal cancer should be seen in a paediatric setting. Usually an appropriate head & cancer specialist, such as a consultant maxillofacial surgery, who also provides a paediatric clinic service can be indentified by your doctor,.*

Mr T Carroll
September 2009