Vaccinations for individuals affected by Fanconi Anaemia

Are there any additional vaccinations that we should give our FA child over and above the normal childhood vaccination schedule?

There are four vaccinations that should be considered for individuals affected by Fanconi Anaemia (FA) over and above the normal childhood vaccination schedule.

The decision concerning the administration and timing of these vaccinations should always be under the supervision of a Fanconi Anaemia specialist, e.g., paediatric haematologist. All such vaccinations have to be repeated, including the childhood vaccines, at an appropriate time point following a successful haematopoietic stem cell transplant as decided by the transplanting paediatric haematologist.

Note that vaccination against chicken pox or hepatitis B has not been specifically recommended in the Fanconi Anemia Research Fund Standards of Care Manual as this publication has been written in a North American context where all children are routinely vaccinated for chicken pox and hepatitis B as part of their normal childhood vaccination programme, unlike in the UK or Ireland.

What is vaccination?

The purpose of vaccination is to introduce the body to some aspect of a particular virus that causes a specific infection so that when the body is subsequently exposed to that virus, the body is already able to activate its defenses by mounting a significant immune response and so preventing infection. If the body has been otherwise exposed to such a virus, vaccination may not be required. This can be checked by testing for antibodies in the blood against the particular virus. The effectiveness of a particular vaccine can also be tested by looking for the subsequent development and persistence of appropriate antibodies for the relevant virus in the blood.

How are these vaccinations given?

All four vaccinations below have to be given by injection. All four vaccinations are considered safe and effective in normal individuals. All four vaccinations may result in a local injection site area of inflammation with some mild discomfort lasting some days.

1. **Chicken Pox Vaccination.** The chicken pox vaccine is a live vaccine using a weaker strain ('Oka' strain) of the varicella zoster virus that causes chicken pox. For children under the age of thirteen, only one injection is required. For individuals over the age of twelve, two injections are required separated by a six week interval. The chicken pox vaccination, as well as sometimes causing a local injection site area of inflammation, may also result in very mild chicken pox type rash. Because the chicken pox vaccine is a live vaccine, it should not be given if an individual is potentially immunosuppressed, e.g., on steroid medication. It should also not be given in severe bone marrow failure. ***The main reason for chicken pox vaccination in FA affected children is that chicken pox infection appears to result in a significant drop in blood counts in at least a third of individuals and in a small minority may result in androgen resistance or the need for haematopoietic stem cell transplantation.*** In normal children, evidence of prior infection by chicken pox is almost universal by ten years of age. However, about a third of children acquire their exposure to chicken pox between the ages of seven and ten. This is a time at which FA affected children are most likely to have low blood counts and be particularly vulnerable to a chicken pox infection. ***Chicken pox infection during this period risks significant complication and the possibility of leading to an urgent unplanned bone marrow transplant with a much reduced time window to search for a matched stem cell donor.***

2. **Hepatitis B Vaccination.** The hepatitis B vaccine is not a live vaccine and instead uses a protein from the Hepatitis B virus to stimulate an immune response. Three separate injections are required over a six month period. ***The main reason for giving the Hepatitis B vaccine is that FA affected individuals will likely require large volumes of blood products, including platelet and red cell transfusions, and will therefore have a small but significant risk of becoming infected with Hepatitis B. Hepatitis B can cause significant liver problems including inflammation of the liver, liver failure, and tumours/cancer of the liver.***

3. **Human Papilloma Virus Vaccination.** The human papilloma virus (HPV) vaccine is not a live vaccine and instead uses a protein from the human papilloma virus to stimulate an immune response. Three separate injections are required over a six month period. ***The main reason for giving the HPV vaccine is because HPV is associated with an increased risk of developing a particular cancer type (squamous cell carcinomas) of the mouth/tongue/tonsil and anogenital areas.*** In the normal population, all cervical cancers in women, and half of mouth/tongue/tonsil, vulval (in women), penile (in men), and anal cancers are caused by HPV. FA affected individuals are particular prone to such mouth/tongue/tonsil and anogenital cancers. Although the role of HPV in FA mouth/tongue/tonsil and anogenital cancers is not conclusive, HPV infection is likely to have a contributory role to the development of at least a proportion of these cancers. HPV infection has been traditionally associated with onset of sexual activity. Hence, HPV vaccination has only been trialed and licensed for girls/women nine years of age and older for the purposes of preventing cervical cancer. However, HPV does occur in younger children independent of any sexual means of transmission. ***Considering the potential risk/benefit profile, HPV vaccination should be given to all FA affected individuals, as young as possible, in both males and females, ideally shortly after diagnosis, and not just left arbitrarily to when the child reaches nine years of age.*** Of the two commercial preparations available, Gardasil is the HPV vaccine that should be given as this covers specifically four of the most common cancer causing HPV subtypes (types 6, 11, 16, and 18) as compared to Cervarix which only covers two (types 16 & 18). ***Note that HPV vaccination is not a substitute for an appropriate cancer screening programme in the FA affected individual. FA affected individuals should have screening examinations every three months for cancer of the***

tongue/mouth/tonsil and also anogenital area, from the age of ten years, by a relevant specialist (e.g., head & neck cancer surgeon for tongue/mouth/tonsil).

4. **Flu Vaccination.** The flu vaccine is not a live vaccine and instead uses proteins from the influenza A and B viruses to stimulate an immune response. The vaccine has to be given each year because of the continuous changing nature of the viruses that cause flu. The vaccine is available in the UK from about September onwards. A thioresal (mercury) free preparation is recommemded. ***An important reason for giving the flu vaccine is that flu in FA affected individuals can significantly reduce blood counts.*** Further information concerning the flu vaccine can be found at http://www.immunisation.nhs.uk/files/flu_factsheet.pdf.

This information sheet has been prepared by Dr Thomas Carroll, an FA affected parent and medical doctor, using the available evidence from the medical literature and also having sought appropriate professional opinions. All decisions concerning vaccination however should only be made following discussion with your own doctor/FA specialist.

1st Dec 2008