

The Lymphatic System, Lymphoedema and Noonan Syndrome

Dr Sahar Mansour of St George's Hospital is leading the research programme which NSA is supporting financially. She introduced us to the lymphatic system, commenting that everyone knows about the cardiovascular system because we learn it at a very early level at school but even a lot of doctors don't know much about the lymphatic system. She explained that the lymphatic system is a really important system with a number of roles:

- it takes back excessive fluid from the peripheries back to the heart so that you don't become swollen
- when you eat fatty food, it's the lymphatics that transport the fats and cause them to be absorbed into the blood system
- it acts as a guard for the body via the immune system, taking up toxins and bacteria to the lymph nodes

It's been known for a long time that people with Noonan Syndrome and CFC can have problems with the lymphatic system. At the specialised lymphoedema clinic which Dr Mansour works at, they have 11 patients with either Noonan Syndrome or CFC with persistent lymphatic problems of whom 10 are male. The age of onset is usually in childhood or early adulthood and can be very young, at the age of 4, but they have had some adults with lymphoedema. They all have swelling of the legs none of them have swelling of the hands or face. Quite often, whether male or female, there is genital swelling - 7 of the 11 have this, which caused quite a lot of distress. 5 have evidence of varicose veins. She went on to explain some of the symptoms which can include

- swelling of the legs
- fluid on the lungs,
- digestive problems with fatty foods,
- an increased risk of infection

and also some of the treatments, including compression, skin care and diet. Sahar explained that there is a lot of work going on to learn more about the condition in people with Noonan Syndrome, some of which NSA is helping to fund. Sahar invited NSA members to help her in the research in two ways:

- by completing a simple questionnaire "Lymphatic Problems in Noonan Syndrome" (which you can find on our website or ask for a copy by emailing pclarke@noonansyndrome.org.uk)
- by those who have a swelling in the legs agreeing to have an MR scan to help the team to improve the imaging of the lymphatics which is one of their key objectives

All the details and forms for these are on the NSA website, where we'll also have a more detailed report on the progress of the research in the near future and a factsheet about Lymphoedema.

