

An overview of her research 2018 in Great Ormond Street Hospital

Presentation Veronica Kinsler

Nevus Outreach Conference July 2018

Report written by Marjolein van Kessel

Notification: this report is based on my personal notes made from the presentation. Though I believe it's quite an accurate report of the presentation, I'm not responsible for misinterpretations or translation errors.

Veronica has done 28 years of study and followed 448 people. There are more people in her registry, but these came in without melanoma). Out of the 448 people 10 developed pediatric melanoma (which is 2%). In the group 10 had fatal melanoma in the brain. All of them had the NRAS mutation.

The mean age was 4.5 yrs., median 2 years of age. Medical literature says 7 yrs. Of age, but a cohort study showed 2 years.

Dr. Kinsler advises single MRI screening of CNS under 6 months of age is the best predictor of all site melanoma risk in childhood. She would like to give treatment before it changes into melanoma. Difficulty is to distinguish between benign and malignant melanoma. Genetic testing is better than histology. Best is to have both tests done to clarify benign or malignant.

Dr. Kinsler doesn't talk about NCM, but melanoma in the brain.

Treatment

4 people have been treated with Trametinib-GSK, and MEK inhibitor. The four children improved, but they did die. She mentions an example of a child already been taken in hospice, was taken out and lived for another 10 weeks. There was rapid and objective improvement. One stayed stable and was asymptomatic for 9 months.

Does it matter what gene you have? 68% is NRAS, 6% is BRAF. Heather's presentation shows BRAF can be the cause.

BRAF is often associated with a distinct multinodular phenotype and histology (shows picture of bulky nevus).

When children have CMN >60 cm PAS, most often is has NRAS mutation.

Past period Veronica has been studying patterns in CMN and vitiligo, another rare condition. She has found patterns that explain the way melanocytes spread in embryonal phase. Veronica showed slides of a disc (early embryo) and the way melanocytes spread and become a trunk of cape nevus.

She sees spontaneous lightening in 112 patients with untreated CMN. L*A*B* is a way to do color measurement by comparing the color of normal skin with colored skin. It shows the color of CMN is significantly related to the background skin tone (dark people get a dark CMN, fair skin people a light CMN). We knew, but now it's proven.

When a baby is born the CMN usually looks very dark. Probably influenced by hormones. Measuring the color is useful after 6 months of age.

Veronica strongly advises not to do curettage. The pigmentation will come back. She showed a picture of a man who had curettage done as a baby on a giant nevus, but a lower satellite was not done. In the picture they had the same color. It proves curettage may seem like it lightens the color of the CMN, but in reality the CMN would have lightened up anyway.

Veronica also showed CMN takes pigment from the body. Three pictures: child with dark hair – CMN dark; child with blond hair: CMN lighter; child with red hair: CMN reddish.