CAFÉ-AU-LAIT MACULE (CALM)

http://www.aocd.org

Café-au-lait macule is also known as CALM, von Recklinghausen spot, or circumscribed café-au-lait hypermelanosis.

A café-au-lait macule (CALM) is a common well-defined flat solitary pigmented lesion that is typically present at birth or early in childhood. It presents as a uniformly light to dark brown area with evenly distributed pigment and sharp borders that typically measures greater than 0.5cm. Its name, café-au-lait macule, comes from its color presentation that resembles coffee with milk. The size of a CALM increases as a child grows and stabilizes in size during adulthood. Adult café-au-lait macule size averages 2-5cm [Range: ≤2mm to >20cm]. They can be found anywhere on the body, except mucous membranes (ie. mouth). The exact cause is unknown when a CALM occurs in isolation, but the resulting pigment is due to an increase in the skin's pigment formation.

Café-au-lait macules can occur in isolation, which are typically benign. A single café-au-lait macule is a normal finding in 10-20% of the population. One to twelve or more CALMs can be seen in individuals, and this can be a normal finding. In fact, 1% of African American children have three Café-au-lait macules. However, when multiple café-au-lait macules are present they have the potential to be associated with genetic diseases. For example, if there are >3 in a Caucasian child or >5 in an African American child then a workup for an associated genetic disease is warranted.

Associated Genetic Diseases:

- Neurofibromatosis
- Legius Syndrome
- McCune Albright Syndrome
- Noonan Syndrome with multiple lentigines (LEOPARD Syndrome)
- Watson Syndrome
- Bloom Syndrome
- Silver-Russell Syndrome

There is no reason to remove or treat a CALM as they are benign and do not become malignant. CALMs are permanent for life and topical treatments are not effective at removal (ie. hydroquinone). Additionally, lasers produce variable results and require varying treatment options and multiple treatments before any noticeable change in the CALM is seen.