



Children's National.

Itch and Skin Care in Patients with NF

A. Yasmine Kirkorian MD

Chief, Division of Dermatology

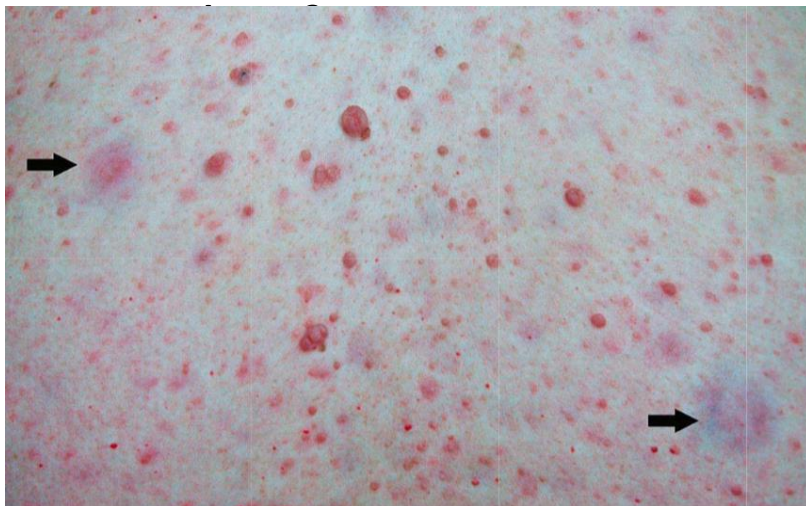
Children's National

Associate Professor of Dermatology & Pediatrics

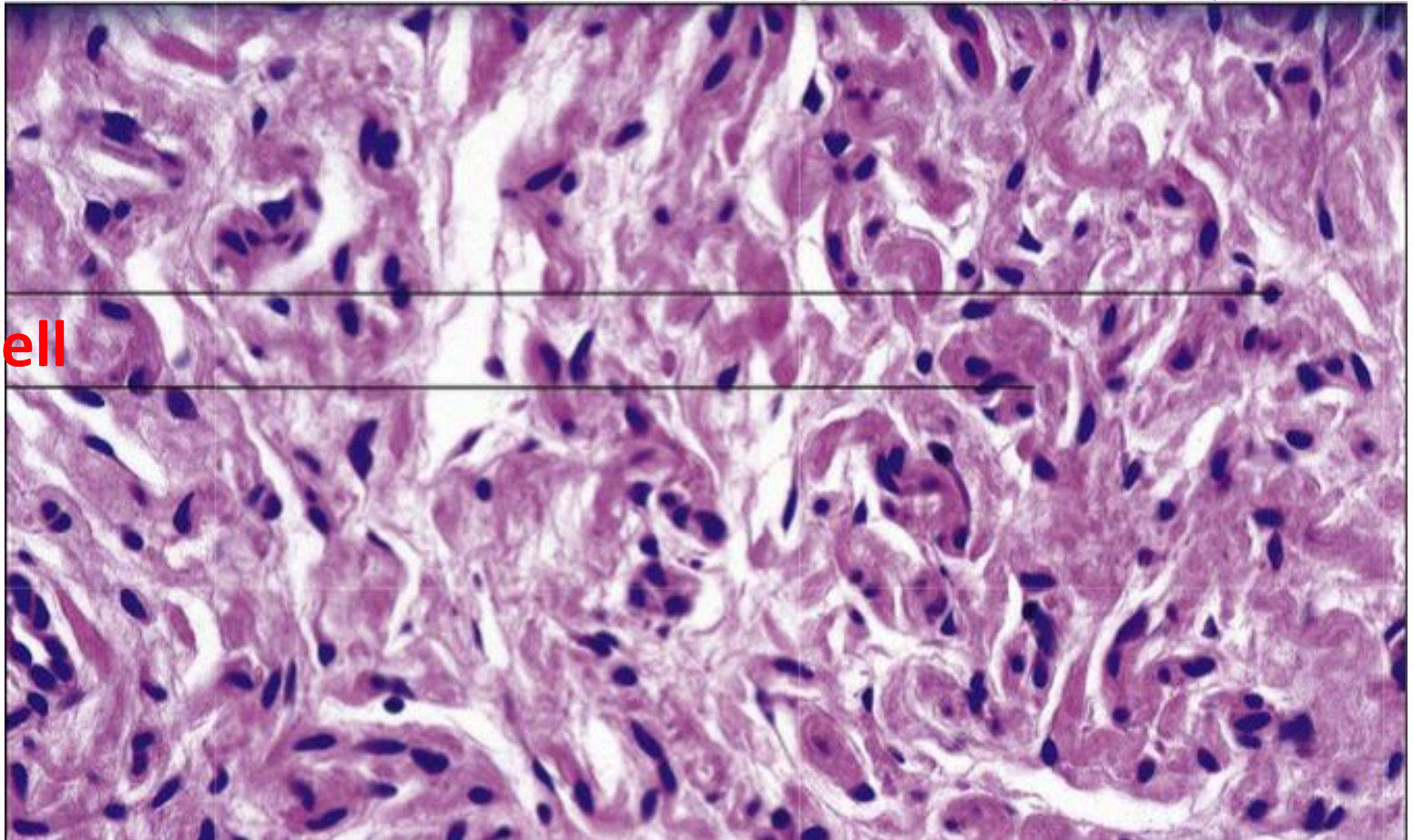
George Washington University School of Medicine and Health Sciences

Objectives

- To review skin findings in NF1
- To discuss general skin care and sun protection
- To review novel treatments for the cutaneous manifestations of NF1



Neurofibroma Histology



Mast Cell

Itch and Pain in NF1

- Studies show that patients with NF1 experience increased skin itching and pain.
- The source of increased itch and pain in patients with NF1 is not clearly known.
 - Sensory nerves are more sensitive?
 - Mast cells in the skin are increased
 - Mast cells cause inflammation, pain and itch
- Itch and pain affect quality of life.

Take Home Point: Good skin care may decrease inflammation and decrease itch and pain.

Skin Care: General Principles

- Moisturize
- Protect from the sun
- Don't smoke

Special Considerations for NF1 Patients:

It is critical to decrease inflammation of the skin by not using harsh skin products and moisturizing daily.

Skin Care: Bathing

- Use fragrance-free, gentle soap
 - Ex: Dove sensitive skin, Cerave, Cetaphil, Aveeno
- Limit soap use to the “stinky” zones
 - Armpits, private parts, feet
- Don’t use wash cloths
- Moisturize the entire body after the shower or bath. Moisturize daily.

Skin Care: Moisturizers

- Thicker is better (brand doesn't matter)
 - Ointments + creams are preferable to lotions
- Fragrance-free is best



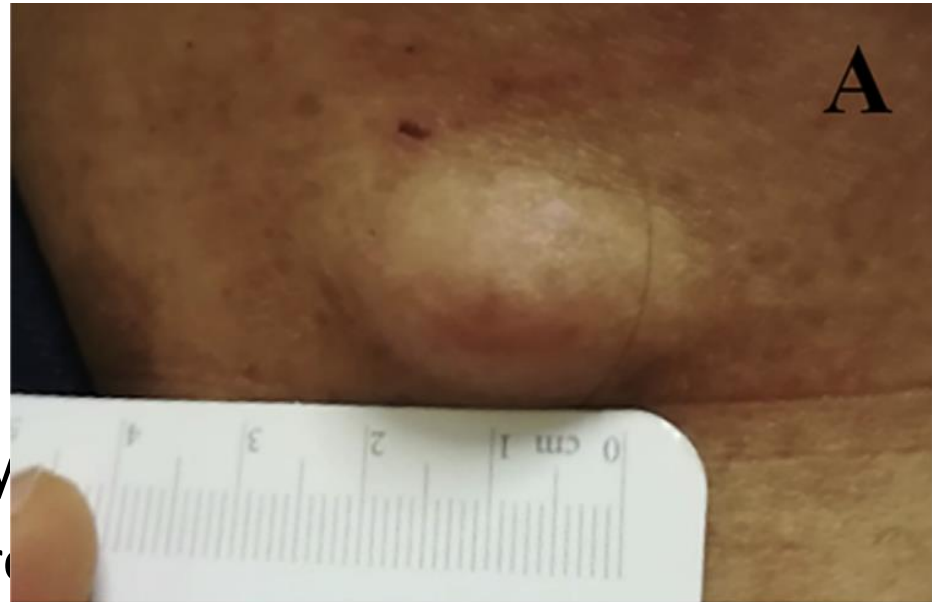
Treatment of Itch

Sarna Sensitive
Anti-histamines

- Non-sedating: Claritin, Zyrtec
- Sedating: Benadryl, Hydroxyzine

Dupilumab?

- Monoclonal antibody blocker of IL-4 and IL-13 for treatment of atopic dermatitis
- Case reports of improvement in pruritus and tumor growth in neurofibromas



Sun Protection: General Principles

- Avoid midday sun (10AM-4PM)
- Wear sun protective clothing
- Seek shade/shade structures
- Use broad-spectrum, >SPF 30 sunscreen daily

Sunscreen



Sunscreen Basics

- Broad Spectrum: blocks UVA + UVB light
- SPF = Sun Protection Factor (UVB light)
 - Ratio of dose of light needed to cause sunburn with sunscreen compared to no sunscreen
- Water Resistance: 40 min, 80 min
- Inorganic: Reflects or scatters light
- Organic: Absorbs light and converts to heat via a chemical reaction

SUNSCREEN AGENTS – MECHANISMS OF ACTION

Sunscreen agents

Inorganic

Organic

Common names

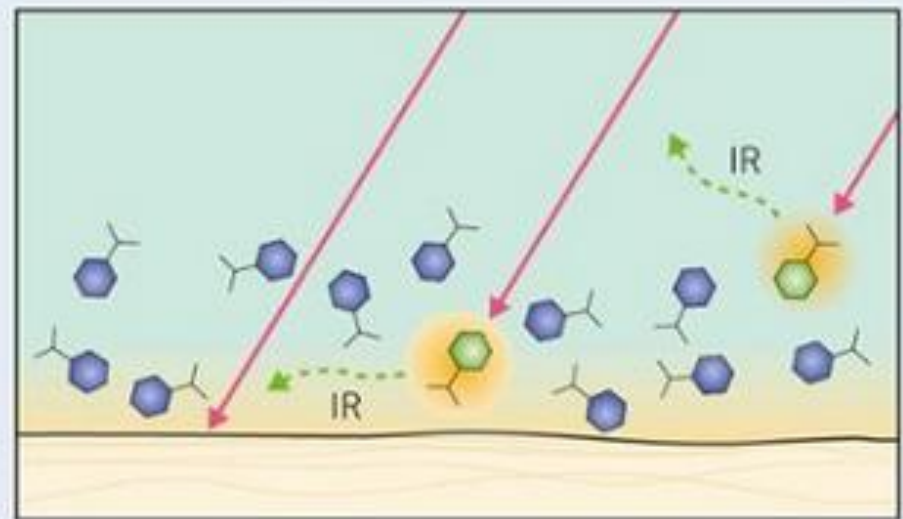
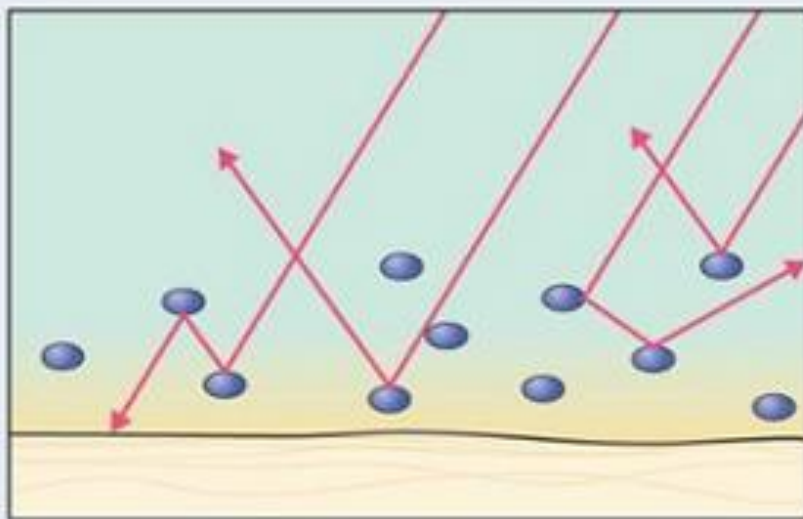
Sunblocks
Physical blockers

Sunscreens
Chemical absorbers

Mechanism of action

Scatter photons

Absorb photons



● Sunblock particle

● Sunscreen molecules

→ UV photons

- - - IR photons

FDA-Regulations of Sunscreen Packaging



Drug Facts	
Active Ingredients	Purpose
Avobenzone 3% Homosalate 10% Octyl methoxycinnamate 7.5%	Sunscreen
Uses	
<ul style="list-style-type: none"> • helps prevent sunburn • if used as directed with other sun protection measures (see Directions), decreases the risk of skin cancer and early skin aging caused by the sun 	
Warnings	
For external use only	
Do not use on damaged or broken skin	
When using this product keep out of eyes. Rinse with water to remove.	
Stop use and ask a doctor if rash occurs	
Keep out of reach of children. If product is swallowed, get medical help or contact a Poison Control Center right away.	
Directions	
<ul style="list-style-type: none"> • apply liberally 15 minutes before sun exposure • reapply: <ul style="list-style-type: none"> • after 40 minutes of swimming or sweating • immediately after towel drying • at least every 2 hours • Sun Protection Measures. Spending time in the sun increases your risk of skin cancer and early skin aging. To decrease this risk, regularly use a sunscreen with a broad spectrum SPF of 15 or higher and other sun protection measures including: <ul style="list-style-type: none"> • limit time in the sun, especially from 10 a.m. – 2 p.m. • wear long-sleeve shirts, pants, hats, and sunglasses • children under 6 months: Ask a doctor 	
Inactive ingredients	
aloe extract, barium sulfate, benzyl alcohol, carbomer, dimethicone, disodium EDTA, jojoba oil, methylparaben, octadecene/MA copolymer, polyglyceryl-3 distearate, phenethyl alcohol, propylparaben, sorbitan laurate, sorbitol, stearic acid, tocopherol (vitamin E), triethanolamine, water	
Other information	
• protect this product from excessive heat and direct sun	
Questions or comments?	
Call toll free 1-800-XXX-XXXX	

How to pick a sunscreen

- Pick the one you will use
- Brand is not important
- Broad-spectrum, at least SPF 30
- Inorganic sunscreens for young children or those with sensitive skin
 - Active ingredient: zinc oxide, titanium dioxide
- Avoid sprays near face due to inhalation risk
- Avoid fragrance if possible

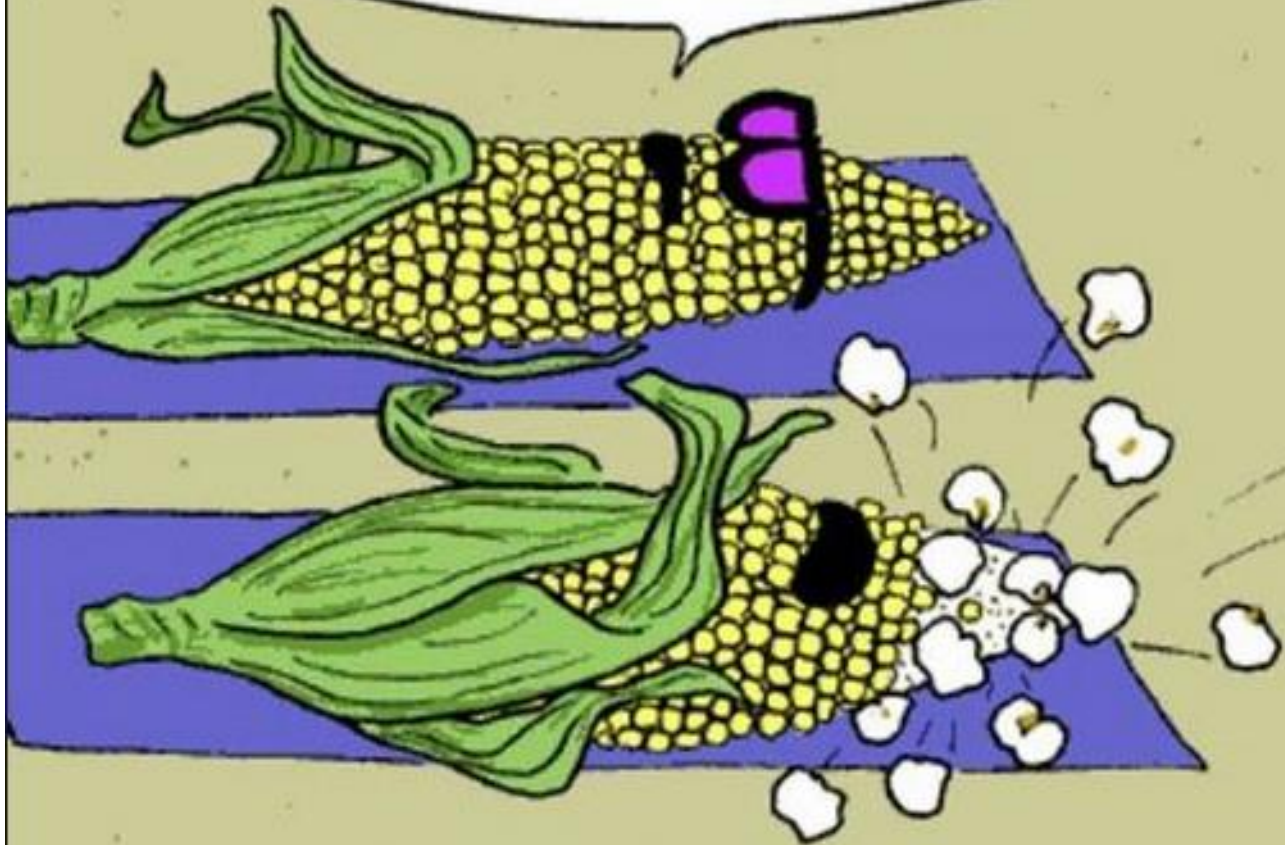
Sunscreen



Sunscreen only works where you use it



I TOLD you to wear sunscreen.



Other sources of sun protection

Before You Go Outside Sunscreen should be your last resort



Wear clothes



Plan around the
sun



Don't get
burned



Wear
sunglasses



Find Shade



Bring Shade



Laser Treatment of Café-au-lait Spots (CALs)



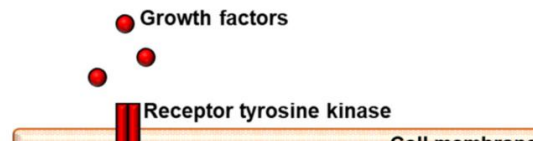
Laser Treatment of CALS

- A variety of lasers have demonstrated efficacy in treating CALM
 - PDL
 - Er:YAG
 - Q-switched Nd:YAG
 - Q-switched Ruby
 - Q-switched Alexandrite
- The durability of pigment improvement is not clear

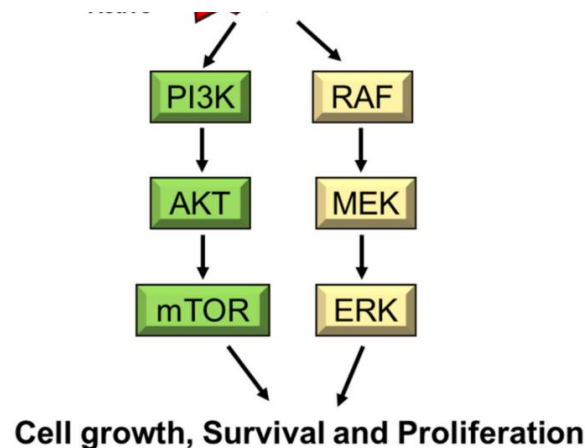
Novel Treatments: mTOR Inhibitors

- NF1 gene deficiency leads to activation molecular pathways called the AKT/mTOR and Raf/MEK/ERK pathways
- mTOR inhibitors (sirolimus and everolimus) block mTOR and have been used to treat plexiform neurofibromas with mixed success
- Topical sirolimus has been used in the treatment of neurofibromas (typical and plexiform)

Novel Treatments: Other Pathway Inhibitors



FDA approves selumetinib for neurofibromatosis type 1 with symptomatic, inoperable plexiform neurofibromas



Summary

- Moisturize every day
- Use sun protective clothing and broad-spectrum sunscreen, SPF 30 daily
- Exciting new treatments are being developed that may treat the skin manifestations of NF1
 - Hopefully certain systemic treatments can be made into topical medications that are easy to apply and safe

Questions?

akirkori@childrensnational.org