
Skin Changes in Proteus Syndrome

Thomas N. Darling, M.D. Ph.D.
Department of Dermatology
Uniformed Services University

Objectives

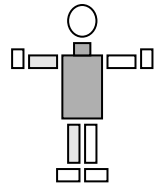
- Identify common skin lesions
 - Anticipate changes in skin lesions
 - Apply principles of good skin care
 - List risks and benefits of potential treatments
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Types of Skin Lesions

- Epidermal nevus
 - Vascular malformations
 - Lipomas
 - Connective tissue nevus
 - Others
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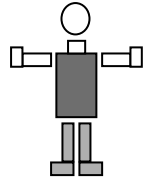
Locations of Skin Lesions

**Epidermal
Nevus**



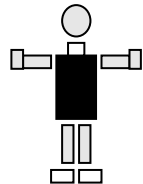
- Torso, Arms, Legs

**Vascular
Malformation**



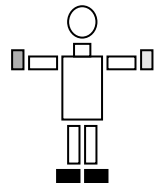
- Torso and Legs

Lipoma



- Torso

**Connective
Tissue
Nevus**



- Feet and Hands

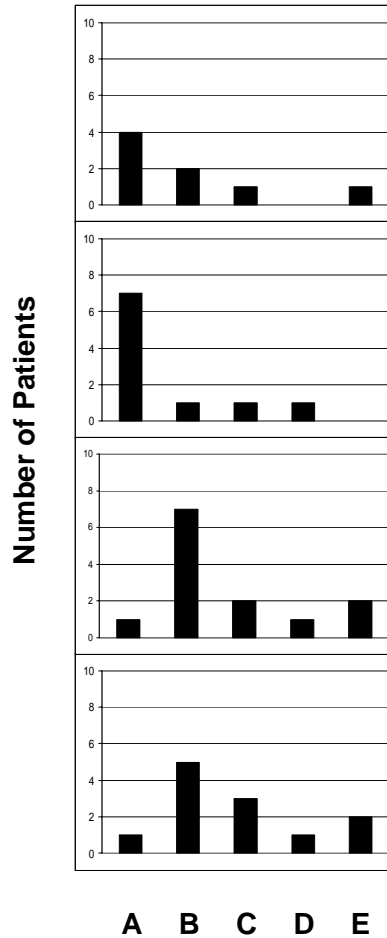
Onset of Skin Lesions

**Epidermal
Nevus**

**Vascular
Malformation**

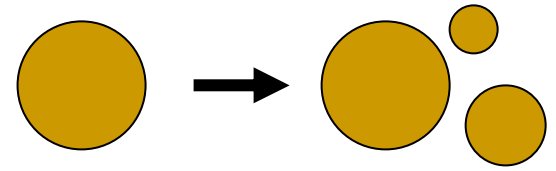
Lipoma

**Connective
Tissue
Nevus**

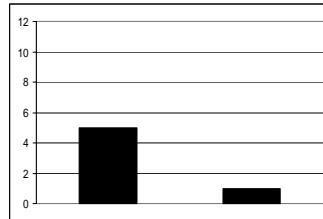


- First year
- First month
- Later in first year
- Later in first 2 years

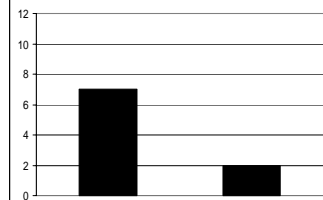
Additional Skin Lesions



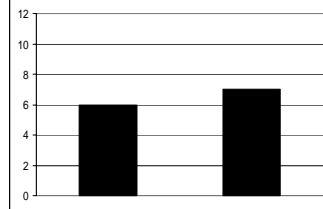
**Epidermal
Nevus**



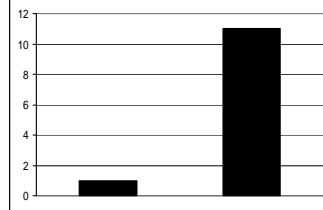
**Vascular
Malformation**



Lipoma



**Connective
Tissue
Nevus**



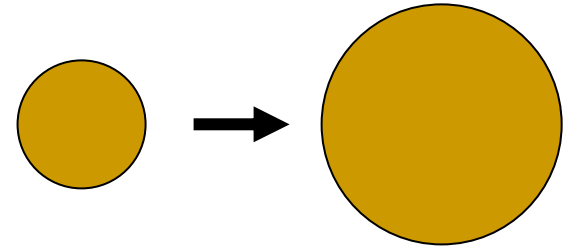
Number of Patients

None

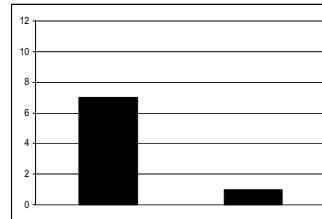
New

- Few new lesions
- Few new lesions
- New lesions later
- New lesions later

Growth of Skin Lesions

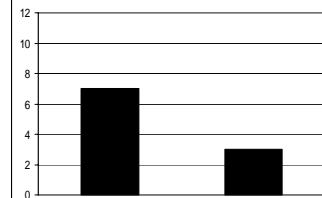


**Epidermal
Nevus**



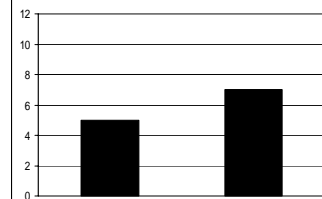
■ Stable

**Vascular
Malformation**



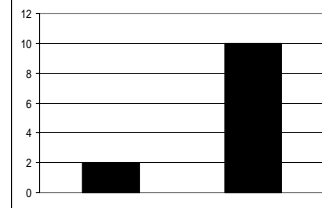
■ Stable

Lipoma



■ Spread

**Connective
Tissue
Nevus**



■ Spread

Stable Spread

Trends

- Early onset, stable size and number
 - Epidermal nevus
 - Vascular malformations (capillary and venous, not lymphatic malformations)

 - Later onset, new lesions, grows
 - Lipomas
 - Connective tissue nevus
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Common Treatment Concerns

- Appearance
 - Impaired function
 - Pain
 - Dry skin
 - Sensitive skin
 - Odor
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Treatment of Dry Skin

- Washing
 - Tepid water
 - Non soap cleanser (e.g. Cetaphil)
 - Bath oil
 - 5 - 10 minutes
 - Shower
 - Pat dry
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Treatment of Dry Skin

■ Moisturizers

- Apply after bathing and 2 – 4 times a day
 - Lotions, ointments
 - Active ingredients
 - Alpha hydroxy acid, salicylic acid, ammonium lactate, urea
 - Patient approval
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Treatment of Sensitive Skin

- Treat dry skin
 - Watch for preservatives, fragrances in moisturizers
 - Avoid irritants
 - Fabrics (wool, rough synthetic clothing)
 - Soaps and detergents (industrial skin cleansers, borax, cleansers with abrasives)
 - Solvents (gasoline, kerosene, turpentine)
 - Acids, alkalis, metal salts, plants
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Causes of Odor

- Moisture
 - Microorganisms
 - Skin folds
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Treatment of Odor

- Keep skin dry
 - Shoes that breathe
 - Cotton socks
 - Change socks
 - Foot powder
 - Hair dryer
 - Gauze wedge
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Treatment of Odor

- Reduce perspiration
 - Antiperspirants
 - Aluminum chloride (Certain Dri)
 - Iontophoresis (Drionic)
 - ? Botox injection



Treatment of Odor

- Reduce bacteria
 - Antimicrobial cleanser
 - Switch brand periodically
 - Foot soaks
 - 10 min, 2-3 times daily
 - Burow's solution (Domeboro), dilute vinegar
 - Clean between skin folds (cotton-tipped applicator)
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Treatment of Odor

■ Prescription

- ❑ Aluminum chloride solution (e.g. Drysol)
 - ❑ Topical antibiotics (e.g. Clindamycin solution)
 - ❑ Treatment of infection (oral antibiotics, topical or oral antifungals)
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Treatment of Skin Lesions

- Different treatments for different types of skin lesions
 - Seek treatment for:
 - Oozing, bleeding
 - Signs of infection (warmth, fever, redness, swelling, pain)
 - Rapid change or growth
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Types of Treatment

- Medical
 - Topical, Oral
 - Gradual and modest improvement
 - Surgical
 - Scalpel, Laser, Dermabrasion, Liposuction
 - Dramatic change
 - Scarring, recurrences
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Choosing the Right Treatment

- Choice of therapy
 - Reasons for treatment
 - Effective
 - Side effects
 - Ease of use
 - Timing of therapy
 - Functional impairment, growth of lesion
 - Age
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Connective Tissue Nevi

- Good skin care
 - Surgery – Caution!
 - Excision (at time of orthopedic surgery)
 - Scars on the sole of foot can be very painful
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Epidermal Nevi

- Medical

- Topical retinoids, keratolytics
- Systemic retinoids

- Surgical

- Excision, shave
 - Laser ablation
 - Dermabrasion
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Vascular Malformations

- Capillary malformation
 - Pulsed-dye lasers
 - Intense pulsed light source
 - Venous malformation
 - Compression stockings
 - Embolization
 - Sclerotherapy
 - Excision
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Vascular Malformations

- Lymphatic malformations
 - Leave undisturbed
 - Excision
 - Sclerotherapy
 - Control infection
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Lipomas

- Surgical
 - Excision
 - Liposuction



Summary

- Children and adults with Proteus syndrome have special needs for skin care – dry skin, sensitive skin, odor
 - Different types of skin lesions are treated in different ways, some medically and some surgically
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Thank you!

- Proteus Syndrome Foundation
 - NIH
 - Patients and Families
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