Photosensitivity disorders in children

Part II

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After completing this learning activity, participants should be able to recognize the pathogenesis and clinical characteristics of hereditary photodermatoses in children; identify dermatoses which may be exacerbated by exposure to UV radiation; and delineate the appropriate diagnostic and management steps for each of these disorders.

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Photosensitivity disorders in children encompass a diverse group of diseases. Some inherited disorders manifest with photosensitivity early in life. Specific extracutaneous association may be the clue to diagnosis in this group of pediatric photodermatoses. Part II of this 2-part review covers hereditary photodermatoses caused by defects in nucleotide excision repair, double strand break repair, or localized or systemic biochemical abnormalities. Diagnosis and management of photoaggravated dermatoses are also discussed. Sun protection strategies are required in all patients with evidence of photosensitivity. Early recognition and prompt diagnosis is essential to minimize the long-term complications associated with inadequate photoprotection. (J Am Acad Dermatol 2012;67:1113.e1—15.)

Key words: children; double-strand break DNA repair; hereditary; nucleotide excision repair; photoaggravated; photodermatoses; photosensitivity; phototesting.

Photosensitivity disorders or photodermatoses refer to skin diseases aggravated by exposure to ultraviolet (UV) or visible radiation. Similar to adults, pediatric photodermatoses encompass a diverse group of diseases and can be classified into 4 main categories: (1) immunologically mediated photodermatoses (IMPs; previously called idiopathic photodermatoses); (2) drug- and chemical-induced photosensitivity; (3) hereditary photodermatoses; and (4) photoaggravated dermatoses. Part II of our review will focus on the last 2 groups.

CAPSULE SUMMARY

- Pediatric photodermatoses encompass a diverse group of diseases.
- Many pediatric photodermatoses are the result of genetic or metabolic defects, and others may indicate underlying systemic disorders.
- Some inherited disorders manifest with photosensitivity, extracutaneous abnormalities and a predisposition to malignancy.
- These genetic diseases can result from defects in DNA repair or localized or systemic biochemical abnormalities.
- The early recognition and prompt diagnosis of pediatric photodermatoses is essential to minimize long-term complications.

HEREDITARY PHOTODERMATOSES

Key point

- **Hereditary photodermatoses are diseases associated with defects in DNA nucleotide excision repair, defects in double strand break repair defects, and defects in biochemical substances**

Some inherited disorders manifest with photosensitivity. These genetic diseases can result from defects in DNA nucleotide excision repair (NER), such as xeroderma pigmentosum (XP), trichothiodystrophy (TTD), and Cockayne syndrome (CS), defects in double strand break repair defects,[1] including Bloom syndrome (BS) and Rothmund—Thomson syndrome (RTS), or defects in biochemical substances, such as Smith-Lemli-Opitz syndrome (SLOS) and Hartnup disease. Asides from cutaneous photosensitivity, specific extracutaneous abnormalities and malignant predisposition are usually seen in these disorders.

Hereditary photodermatoses caused by defects in nucleotide excision repair

XP, TTD, and CS, including cerebro-oculo-facio-skeletal (COFS) syndrome and UV-sensitive syndrome (UV[S]S) are rare autosomal recessive neurocutaneous disorders caused by defects in NER. The NER pathway serves to repair DNA that is damaged by UV radiation, chemical carcinogens, and other non—UV-induced oxidative damage.[1] There are at least 28 proteins in the NER pathway. Three of the proteins are also part of the 10-protein complex of the transcription/DNA repair factor IIH (TFIIH). The NER pathway is therefore closely linked to transcription.

NER also has a role in normal human development, including the development of the nervous system. There is a complex relationship between the

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