

DERM Dx

A girl born at 41 weeks' gestation was apneic upon delivery and had a rash from head to toe. She was intubated and sent to the NICU. Skin exam showed erythematous papules with purpura, crusting, and ulceration that involved the face, trunk, arms, legs, palms, and soles. The mother had a history of spontaneous abortion, but was up to date on vaccinations and her screening exams were normal. What's your diagnosis?



SAN DIEGO — The most aggressive variant of Langerhans cell histiocytosis, Letterer-Siwe disease is characterized by cutaneous involvement, organomegaly, and thrombocytopenia, Joseph P. Janik, M.D., said during a poster session at the annual meeting of the Society for Pediatric Dermatology.

The patient was thrombocytopenic and had an enlarged spleen and liver, but there were no lung abnormalities. The results of two shave biopsies confirmed the diagnosis, said Dr. Janik, the lead author of the case report, of the department of dermatology at the University of New Mexico, Albuquerque.

Few controlled studies exist for the treatment of the Letterer-Siwe variant of Langerhans cell histiocytosis, let alone the congenital form. PUVA and topical nitrogen mustard seem to be most effective for the skin lesions while vinblastine or etoposide have been used for the systemic manifestations (J. Pediatr. 1991; 119:317-21). For nonresponders, a

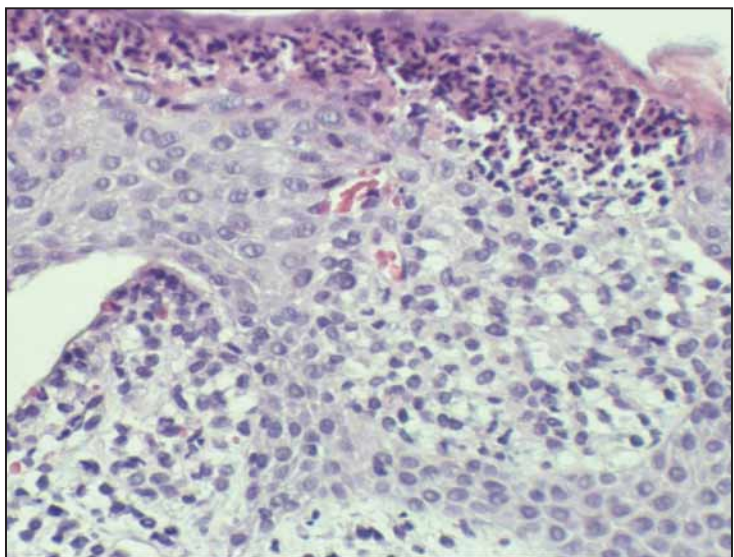
combination chemotherapy can be tried.

The response rate to monotherapy can reach 90%, but the overall outcome is usually fatal, especially in the congenital form of Letterer-Siwe (Cancer 1988;62:2528-31). At press time, this patient is alive at the age of 21 months, after a year of treatment with vinblastine, prednisone, and 6-mercaptopurine.

Dr. Janik discussed another case of the Letterer-Siwe variant of congenital Langerhans cell histiocytosis in a baby born at 28 weeks' gestation. Apgar scores for the baby, who was delivered via emergent C-section, were 2 at 1 minute, 1 at 5 minutes, and 1 at 10 minutes. Resuscitation attempts were unsuccessful, and the baby expired 25 minutes after birth.

Ran H. Bang, M.D., Charles H. Palmer, M.D., E. Ben Smith, M.D., and R. Steven Padilla, M.D., all of University of New Mexico, assisted with the case reports.

—Doug Brunk



PHOTOS COURTESY DR. JOSEPH P. JANIK

Low-Dose Flutamide May Help Treat Female Refractory Acne

BY JEFF EVANS
Senior Writer

QUEBEC CITY — Dosages of the androgen receptor blocker flutamide at 125 mg/day appear to be effective in treating acne in women who have not responded to other medications, James C. Shaw, M.D., reported at the annual conference of the Canadian Dermatology Association.

Flutamide has been reported to be effective in treating acne at doses of 500 mg/day and 250 mg/day. But flutamide has not been used as widely as other androgen receptor blockers such as cyproterone acetate and spironolactone because of an incidence of hepatotoxicity ranging from 1% to 5% at dosages greater than 500 mg/day and in isolated cases at 250 mg/day, according to Dr. Shaw of the division of dermatology at the University of Toronto.

In a review of 32 consecutive women aged 14-

51 years who received a prescription for flutamide at 125 mg/day, 17 of the 21 patients who returned for follow-up visits had marked improvement of their acne. The length of treatment in the 21 patients ranged from about 2 months to 21 months, Dr. Shaw and his associates wrote on a poster.

Overall, five women discontinued treatment because of emotional lability (two patients), minor GI distress (two), or a slightly elevated level of alanine aminotransferase (one). With the exception of four patients, none the women had responded adequately to or had tolerated other therapies.

Dr. Shaw said that he regularly conducts liver function tests. Flutamide has been associated with developmental abnormalities in exposed fetal rats, so all patients must be advised about contraceptive use during treatment. The drug is most often used to treat prostate cancer in men and hirsutism in women.



This patient's acne was refractory to treatment with isotretinoin.



Her acne responded to 8 months of flutamide 125 mg/day plus oral contraceptives.

PHOTOS COURTESY DR. JAMES C. SHAW

Tips for Improving Teenagers' Adherence to Acne Treatment

BY ROBERT FINN
San Francisco Bureau

SAN FRANCISCO — Lack of treatment efficacy is only one of the reasons that acne therapy often fails in teenagers, Lee T. Zane, M.D., reported at a meeting on clinical pediatrics sponsored by the University of California, San Francisco.

Poor adherence is also a factor, said Dr. Zane of the university. And poor adherence can stem from several things. The treatment regimen may be too complex, or patients may stop treatment prematurely when they don't see quick results. Adverse effects of therapy often lead to poor adherence too. In some cases, the patient may have conceptual opposition to certain modes of therapy.

Dr. Zane offered these tips to optimize adherence:

► **Simplify the treatment regimen.** Use fewer agents or combination agents. Combina-

tions of clindamycin and benzoyl peroxide are available now. Coming soon there may be retinoids plus antibiotics and retinoids plus benzoyl peroxide.

► **Minimize the adverse effects.** Because benzoyl peroxide can be especially irritating, maximize retinoid therapy before maximizing benzoyl peroxide.

With retinoids and benzoyl peroxide, advise patients that a pea-sized dollop should be enough to cover the face. Using more will not improve the result, but will increase irritation.

Advise them to apply topical agents to dry skin, waiting 20-30 minutes after washing.

► **Set realistic expectations.** Tell patients to try a therapy for at least 2 full months before deciding whether it's effective.

"I'm going for a slow and steady course of therapy followed by long-term maintenance," Dr. Zane said. "It's not a quick cure. It's really about management of a chronic condition and prevention.

► **Remember that your clinical assessment of disease severity may differ dramatically from the patient's assessment.** A single pustule on otherwise porcelain skin may be more damaging psychologically than widespread disease.

► **Peer opinion is often far more compelling than scientific data.** The physician may be focusing on the patient's face, but if his friends are making fun of the acne on his back, he'll be more concerned about that.

► **Warn patients about behaviors that worsen acne.** Physicians have long advised against picking or squeezing, but they also should point out that scrubbing or exfoliating can promote the formation of comedones. Rubbing the skin during sports or other activities also can promote acne.

Finally, some medications can exacerbate symptoms. These include lithium, topical and oral corticosteroids, and androgenic steroids.