ly proven chronic active hepatitis with anti-smooth muscle antibody. The others did not possess anti-smooth muscle or anti-mitochondrial antibodies.

It has been proposed that immune complexes are formed in the intestine in coeliac disease (9). We did not find any immune complexes in the DH patients with hepatic injury, although the present techniques may be insufficiently sensitive to detect them. Hence it seems difficult to postulate that these are the mediators.

Our results contrast with those of Stone & Goodwin (11) who showed a 50% incidence of obstructive jaundice in patients taking dapsone for acne. None of our 30 patients taking dapsone had an elevation of the alkaline phosphatase level. This may be due to the lower doses used in DH (50–150 mg daily), compared with 150–200 mg daily for acne patients. The incidence of haemolytic anaemia was approximately the same in both series, viz. 1 out of 6, or 17%. It is unlikely that the abnormalities of AST and GGTP observed in our patients were due to the dapsone, as in several cases they returned to normal without any reduction in their dapsone dosage.

Despite the difficulty of adhering to a GFD the lower incidence of abnormal liver function in patients with DH on a GFD in contrast to a normal diet is yet another reason for advising this treatment, where it seems practical.

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A Spontaneously Healing Collodion Baby: A Light and Electron Microscopical Study[1]

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Abstract. Skin biopsies from a collodion baby, spontaneously healing at the end of the third month, were taken on the 1st and 15th day after delivery and examined by light and electron microscopy. The microscopical features observed were different from those known to occur in collodion babies evolving into lamellar ichthyosis and may

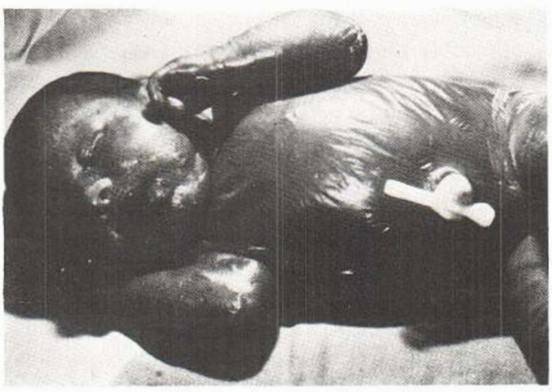

Fig. 1. Clinical appearance of the propositus on the first day after delivery: typical collodion baby.

[1] Presented at the 7th European Meeting of the Society for Cutaneous Ultrastructure Research, Vienna, May 9–10. 1980.

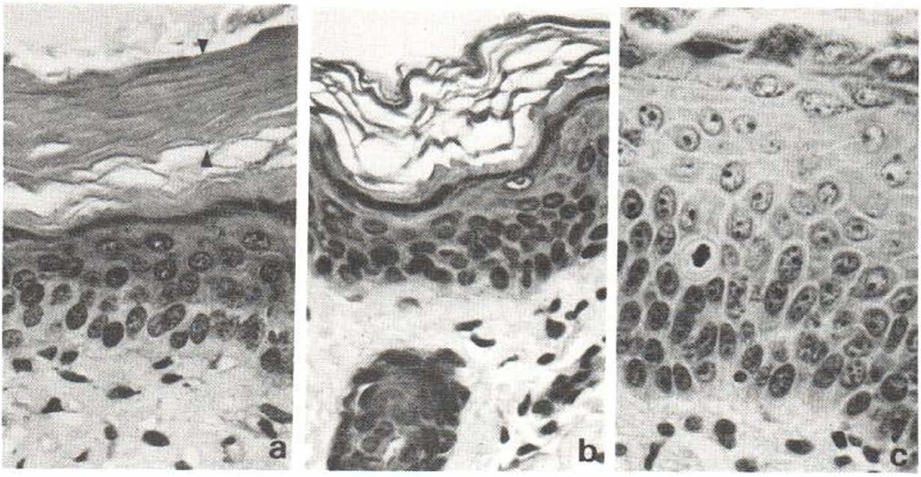

Fig. 2. Light microscopy of neonatal skin (PAS, ×400). (*a*) Propositus, first-day biopsy: thickened compact stratum corneum, the upper 2/3 are PAS-positive (▼▲); normal epidermis. (*b*) Propositus, 15th day biopsy: normal stratum corneum and epidermis. (*c*) For comparison, 15th day biopsy of a collodion baby later developing lamellar ichthyosis: marked hyperplasia of the epidermis, mitotic figures frequently seen.

contribute to a more precise, early diagnosis and prognosis of this heterogeneous neonatal syndrome.

Key words: Collodion baby; Ichthyosis; Lamellar ichthyosis of the newborn

The collodion baby represents a rare heterogeneous syndrome of the newborn, which is characterized by a collodion-like membrane covering the entire body surface. The great majority of the surviving

Table I. *Light and electron microscopy of skin from a spontaneously healing collodion baby in comparison with collodion babies evolving into lamellar ichthyosis*

	Spontaneously healing collodion baby		Collodion baby evolving into lamellar ichthyosis	
	1st day biopsy	15th day biopsy	1st day biopsy	15th day biopsy
Light microscopy				
Stratum corneum	Thickened Lamellar, compact Upper 2/3: PAS-positive	Normal thickness Basket weave Some PAS-positive lamellae on surface	Thickened Lamellar, compact Upper 2/3: PAS-positive	Thickened Lammellar, compact Some PAS-positive lamellae on surface
Epidermis	Normal	Normal	Normal	Hyperplasia
Mitotic figures	Normal	Normal	Normal	Increased
Electron microscopy				
Stratum corneum	Thickened Normally arranged lamellar stratum corneum cells	Normal thickness Normal morphology	Thickened, compact Upper 2/3: large convoluted horny cells, keratinization of irregular density, remnants of nuclei and organelles	Thinner than on 1st day Lamellar horny cells with numerous vacuoles
Epidermis	Normal architecture	Normal architecture	Upper-keratinocytes less flattened than normal, rich in ribosomes and mitochondria Intercellular oedema	Many transitional cells marked hyperplasia
Keratohyalin	Normal	Normal	Decreased, present in 1–2 layers	Decreased, present in 3–4 layers
Tonofibrils	Normal	Normal	Decreased	Decreased
Odland bodies	Normal	Normal	Normal	Normal

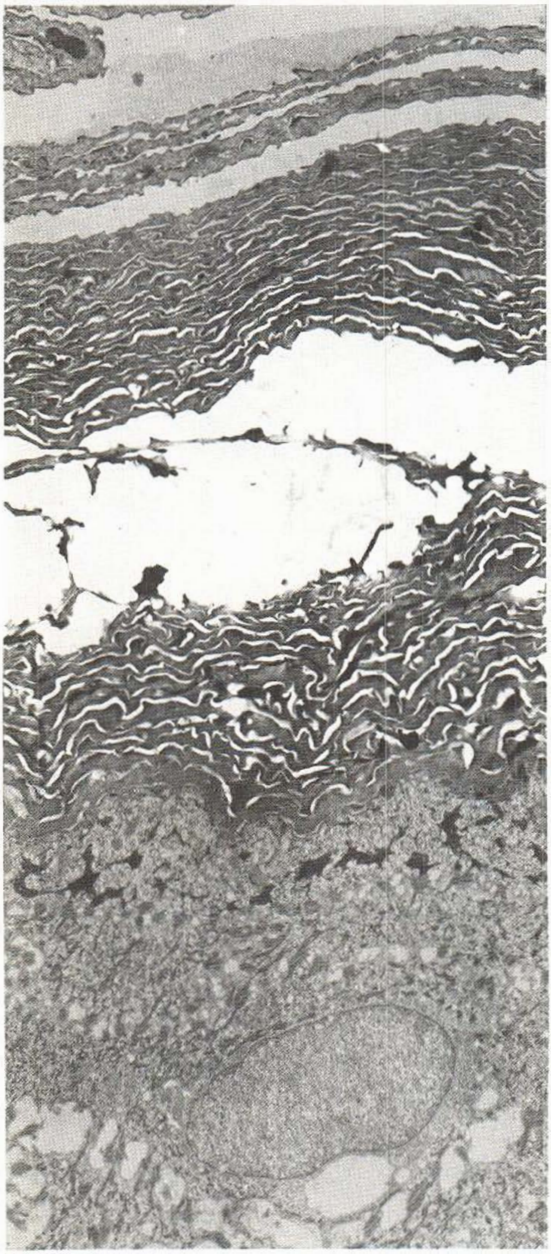

Fig. 3. Electron microscopy of collodion skin from the propositus (first-day biopsy): stratum corneum hyperkeratotic but of normal structure, intercellular oedema of the normal epidermis (×5 000).

cases evolve into lamellar ichthyosis (non-bullous ichthyosiform erythroderma). Occasionally a different outcome has been reported, such as autosomal dominant and X-linked ichthyosis vulgaris, ichthyosis linearis circumflexa, or other rare ichthyosiform syndromes (2). Recovery has been observed in 3.5% of the published cases (2); such cases probably correspond to what has been called in a rather equivocal way "lamellar ichthyosis of the newborn" (3). At present, reliable criteria for evaluating the later outcome of a collodion baby are not available.

The sequential light and electron microscopical alterations of neonatal collodion skin have only been studied in cases of lamellar ichthyosis (1). We now report the light and electron microscopical features of a clinically typical collodion baby, whose skin condition cleared during the first 3 months of post-natal development.

CASE REPORT

B. G., a boy, was a first child, born after a pregnancy of 9 months. The birth weight was 3 390 g. During pregnancy the mother had been treated for a serological toxoplasmosis. At birth the patient's entire body was covered with a thin, shining, collodion-like membrane on a red oedematous skin (Fig. 1). The collodion membrane soon became wrinkled and fissured in flexural areas. A slight ectropion of the eyelids was also noted. The collodion membrane shed during the first month; the skin became moderately scaly and cleared completely by the end of the third month. The general neonatal development was normal, without complications.

The parents of B. G. were second-degree cousins. A first-degree cousin of the maternal grandmother of B. G., a girl, reportedly had had a similar, transient neonatal ichthyosiform dermatosis.

Biopsy samples were taken from B. G. 7 hours after birth and on the 15th day from symmetrical areas of the anterior aspects of the arms. The two biopsy specimens were processed according to routine methods for light and electron microscopy.

The results of the light and electron microscopical study, illustrated by Figs. 2 and 3, are reported in Table I and compared with the microscopical alterations previously found in collodion babies subsequently developing lamellar ichthyosis (1).

COMMENT

Our results suggest that microscopical examination of neonatal skin could become a valuable tool for establishing a more precise early diagnosis and prognosis of a collodion baby. At least, it appeared to be possible to distinguish between the type associated with lamellar ichthyosis and the spontaneously healing variety. At the age of 15 days, at a stage where the clinical picture is still dominated by collodion-like lamellae, the light microscopical features of the underlying epidermis were already

those of the later, definitive state of the skin. Furthermore, the marked ultrastructural abnormality of the upper stratum corneum, found in neonatal collodion skin of lamellar ichthyosis, was not observed in the self-healing collodion baby. These results should encourage early microscopic examination and follow-up of all types of collodion babies, such data being needed to elaborate reliable, early diagnostic and prognostic criteria.

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Stevens-Johnson Syndrome Associated with Intrahepatic Cholestasis and Respiratory Disease: A Case Report

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Abstract. In an 8-year-old boy, the Stevens-Johnson syndrome (SJS) was associated with protracted intrahepatic cholestasis and ultimately fatal respiratory disease. No precipitating factors of SJS were identified, but a *Mycoplasma pneumoniae* infection was suspected. The appearance of jaundice on the seventh day of illness was preceded by the prescription of erythromycin ethyl succinate, but intrahepatic cholestasis has never before been associated with this derivative of erythromycin. It is suggested that there may exist a subtype of SJS with severe hepato-pulmonary pathology.

Key words: Stevens-Johnson syndrome; Intrahepatic cholestasis; Bronchiolitis; Pneumomediastinum; Erythromycin ethyl succinate

Stevens-Johnson syndrome (SJS) is an acute clinical entity which encompasses fever, exanthema, and inflammation of the orifices of the body (10). SJS may involve internal organs, especially the respiratory and the alimentary tracts (1). There are only occasional reports on liver pathology in association with SJS. We report a unique case of SJS associated with intrahepatic cholestasis and fatal respiratory disease.

CASE REPORT

An 8-year-old boy had suffered from a scaling dermatitis since early childhood. The patient had no record of adverse drug reactions or earlier treatment with erythromycin.

On March 5, 1979, the patient fell ill, with a temperature rising to 40°C. During the next 24 hours he developed an unproductive cough, a red non-vesicular exanthema, conjunctivitis, and erosions of the lips and mouth, glans penis and anus. He was admitted on March 8, the temperature then being 40.4°C, and a generalized purple macular exanthema was found with isolated bullae on the cheeks and extensive ulcerous lesions of the orifices of the body, which led to the immediate diagnosis of SJS.

Examinations on admission. The hemoglobin was 13.7 g/dl, the erythrocyte sedimentation rate 24 mm/h, the leukocyte count 7800/μl with normal distribution. The chest X-ray appeared normal except from subsegmental atelectases of the left inferior lobe.

Treatment. Mycoplasma infections were prevalent in our area at that time, and an underlying infection with this microorganism was suspected (8, 10). Erythromycin ethyl succinate (Abboticin®, Abbott), 300 mg four times a day, was started on March 8 and given for 11 days. The patient's weight was 27 kg.

Topical treatment was given, i.e. prednisolone and chloramphenicol for the eyes, diluted potassium permanganate for the mouth, ointment with silver nitrate for the lips, and mucilago borica for the genital and anal erosions.

Clinical course. The general condition of the patient improved parallel with lytic defervescence within 2 weeks of admission, and the mucocutaneous lesions gradually healed. Mild respiratory discomfort with a productive cough persisted. Jaundice was observed on the fourth day of hospitalization, and it took a protracted course (further details below and in Fig. 1). On April 5, the liver disease prompted a therapeutic trial with prednisone, 40 mg per day, withdrawn over a period of 12 days, but no immediate response was observed.

On April 8, the patient developed stridor with pneumomediastinum and interstitial emphysema, which