could give no prevalence of HP among dentists in the epidemiological sense of the word, but still the survey indicates the existence of an occupational disease of an order of magnitude, which makes further study relevant.

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REFERENCES

The CHILD-Syndrome—Congenital Hemidysplasia with Ichthyosiform Erythroderma and Limb Defects. A Case Report
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A presentation of a 3-year-old girl with a congenital hemidysplasia of the left side together with ichthyosiform erythroderma, the so-called CHILD-syndrome. A short review of the syndrome is given. Key words: X-linked dominant gene-defect, Lyon-effect. (Received September 9, 1983.)

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In 1980 Happle et al. (1) proposed the term "CHILD-syndrome" for what was previously named "congenital unilateral ichthyosiform erythroderma". The reason for introducing the new name was to give a better description of the syndrome and to focus on new genetic aspects.

In 1982 Happle (2) had collected a total number of 25 published cases—all but one of female sex. The present paper describes another case of the CHILD-syndrome, probably a mutant.

CASE REPORT
A 3-year-old girl was admitted for the first time to our clinic at the age of three months. The family history was negative and her parents not related. A 6-year-old sister was without symptoms. Her birth
Fig. 1. The skin eruptions are localized only to the left side of the body and extremities.

Fig. 2. The shortness of the left leg is clearly seen.

weight was 2.790 g and her length 48 cm. The mother had no history of abortions, and the present pregnancy and birth was without complications. At birth atrophy of the left-sided extremities and syndactyly of the fourth and the fifth finger was observed. When examined by us we found the skin of the left side of the body and the left-sided extremities hyperkeratotic and erythrodermic and diagnosed the case as a "congenital unilateral ichthyosiform erythroderma" or CHILD-syndrome (Fig. 1). The skin was primarily treated with neutral ointments containing salicylic acid 1%.

A certain improvement was observed during the first few years. However, at the age of three exacerbation took place, which motivated introduction of systemic treatment with Tigason® (Etretinate). All skin symptoms were still strictly located to the left side of the body and the left leg was found to be 6 cm shorter than the right one (Fig. 2). At present it is still too early to estimate the results of this treatment.

Histopathology
In a punch-biopsy from the left axillary region a thickened epidermis with acanthosis and a lamellar hyperkeratosis with prominent parakeratotic foci was found. The finding was most pronounced over the elongated papillae. The keratinization superficial to the rete ridges was orthokeratotic, and there was a thickened granular layer. No ballooning was observed. The acanthotic epidermis contained several dyskeratotic cells, and in the basal layer there was spongiosis. The papillary dermis revealed slight oedema and diffuse infiltration of lymphocytes. The histological changes were similar to those previously described for recessive, congenital ichthyosiform erythroderma.
DISCUSSION

The inheritance of this abnormality has previously been described as an autosomal recessive transmission. Happle et al. (1) proposed an X-linked dominant gene-defect lethal in the hemizygote male foetus.

Furthermore, in 1982 Happle (2) proposed an explanation for the unilateral distribution of the defects, referring to the so-called “Lyon-effect” (3, 4). The hypothesis is, that during the first weeks of foetal life an inactivation of one of the X-chromosomes in all somatic female cells will occur. This may happen at different stages. If it occurs at an early stage, half of the body cells will receive the normal X-chromosome from the father, and the other half the defect chromosome from the mother. Thus, in one half of the body the cells will have the normal gene from the father and in the other half the defect gene from the mother.

The case of the CHILD-syndrome published here is most likely to be a mutant. In her family there are no other cases or abnormalities.

Her skin problems are obviously grave, but the most serious abnormality seems to be the retarded growth of her left-sided extremities.

REFERENCES

Improvement of Progressive Systemic Sclerosis (PSS) with Estriol Treatment

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Two female patients suffering from progressive systemic sclerosis (PSS) of generalized type were treated with estriol for 10 months. Although complete cure was not achieved, considerable beneficial effects were observed in these patients, but no serious side effects. Thus, skin softening was observed on all involved parts of these patients, which was accompanied by increased mobility of large or medium-sized joints and improved pigmentation and cyanotic redness on fingers and extremities. However, no apparent improvement of internal organ involvement could be detected. Histopathologically, a drastic reduction of homogenization of collagen bundles in the dermis of affected forearms was seen in these patients following estriol treatment. To our knowledge this is the first report of estriol therapy for PSS. Key words: Homogenization of collagen bundles. (Received June 30, 1983.)

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Progressive systemic sclerosis (PSS) is thought to be a disorder of connective tissue which affects not only the skin but also a variety of internal organs. Its pathogenesis is still unknown (1, 2). Although a few drugs are known to affect the course of this disease (1, 3), a complete cure is as a rule not expected (4). The following facts happened to make us consider estriol therapy for PSS. Firstly, PSS affects primarily women (4). Secondly, pregnancy sometimes shows a favorable effect on PSS (5, 6, 7). Thirdly, estriol comprises the greater part of urinary estrogen in late pregnancy (8). Fourthly, estriol has a softening effect on the cervix uteri during parturition. Lastly, estriol inhibits osteoporosis (9), which is not only an aging effect but also one of the clinical manifestations of PSS. To our knowledge, estriol therapy for PSS has not been reported before. We describe here the results of such treatment.

CASE REPORTS

Two patients with PSS who gave prior informed consent were treated with estriol. Their diagnosis was confirmed by clinical and histopathological investigations. In these patients, cutaneous involvement was found on the trunk and extremities (generalized type). Examinations made included erythrocyte sedimentation rate (ESR), urinalysis, blood count, blood chemistry, serological tests, X-ray examination of chest, gastrointestinal tract and hands, pulmonary function tests, electrocardiogram, sialography and so on.

According to the American Rheumatism Association's proposal of "proximal scleroderma" as the sole major criterion of systemic sclerosis, whose sclerodermatous cutaneous changes were defined as tightness, thickening and non-pitting induration (10), we made much of inspection and palpation when assessing cutaneous involvement. e.g. on pigmentation, wrinkle formation, the degree of skin being lifted by fingers and so on. As PSS usually begins with edema or swelling of fingers and progresses to hidebound changes (1, 11), we classified cutaneous involvement into five stages as follows: stage 0 =