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We hope that making available the relevant information on Pachyonychia Congenita will be a means of furthering research to find effective therapies and a cure for PC.
Pachyonychia Congenita with Epi-
dermal Cysts and Teeth at Birth: 4th
Generation.—Dr. F. S. Bissen (for Dr
E. J. Moynahan).

A girl aged 4 months (IV 3), her mother
(III 3 the original proband), her maternal
aunt (III 1) and her maternal cousin
(IV 1).

History.—At birth she (IV 3) was found
to have three "gristly teeth," which were
lower incisors. Her nails were normal at
birth, but at the age of 10 days "yellow
spots" were observed on the nail-plates.
From this time onwards, until she was
4 weeks old, the nails became markedly
thickened.

Family history.—(Fig. 1) Her father (not
shown on the pedigree) is alive and well.
Her mother (III 3) belongs to a large
family with the names Morgan-Williams-
Brown who have pachyonychia congenita,
slow growing hair, hyperkeratosis of some
areas and epidermal cysts in the adults.
In most of the affected individuals, teeth
have been noticed at birth.

On examination.—She was a healthy
alert infant, with 3 loose teeth (lower
incisors). All the nails were thickened
and in particular those of the big toes.
The hair was fine, and she had a friction
alopecia in the occipital and parietal
regions. There was no evidence of hyper-
keratosis or of leukokeratosis oris. Her
mother (III 3) shows the fully established
condition with marked epidermal cyst
formation, mainly in the neck area. Her
aunt (III 1) also shows the fully estab-
lished picture with very marked epidermal
cyst formation, again mainly in the neck
area. Her cousin (IV 1) has sparse hair,
thickened nails, hyperkeratosis over the
pre-patellar and elbow areas and early
cyst formation in the axillae. None of
the members present display any leuko-
keratosis oris.

Comment.—The family of which this
girl is a member was first reported by
Jackson and Lawler (1951). Shrank
(1968) reported another sibling (IV 1).
The term pachyonychia congenita was
first used by Jadassohn and Lewando-
wski (1906) when they described a girl aged 15
who showed thickened nails, hyperker-
atosis around the nose and shin with some
vasicle formation, hyperhidrosis of the
nose and callousness of the palms of the
hands and soles of the feet. Her elbows
and knees showed a somewhat follicular
hyperkeratosis. A thick grey cover was
noted on the tongue and the edges showed
white streaks.

This baby and her family were presented
for the following reasons: (i) She was
brought along soon after birth to show the
loose "gristly" teeth, which will probably
fall out soon, and to show that she has
already developed thickened nails, starting
with "yellow spots" at the age of 10 days.
She does not show any hyperkeratosis or
leukokeratosis oris at this age. (ii) The
family also demonstrates the unfolding of
the natural history of this disorder,
starting with abnormal teeth at birth, the
relatively early development of cysts in
both axillae (IV 1) and finally the fully
established picture of nail thickening,
sparse hair, hyperkeratosis and multiple
epidermal cysts in the adults. The teeth
of IV 1 are serrated. (iii) It is suggested
that the present condition must be
separated from that described by Jadas-
sohn and Lewandowski (1906). No men-
tion is made in their paper that the girl
had teeth at birth or epidermal cysts
when she was an adolescent. At the age
of 15 she had not developed the latter.
None of the members in this family show
leukokeratosis oris, which was a striking
feature in the patient described by Jadas-

"Cutiis Nodu-
sica. Dtsch.

"Amyloidosis
(1969, p. 304.

"Arch. Haut.

Beitrag zur

arch. Klin.

repotted in the

1969, p. 304.

Fig. 1.
sohn and Lewandowski. In this family
the gene behaves as an autosomal dominant
trait and breeds true. It is interesting to
note that a male member (III 2) died
from leukaemia. The significance of this
is not known. Soderquist and Reed
(1968) recently reported 3 individuals in
3 successive generations of one family
who showed almost identical characteris-
tics to the family presented today. The
possibility that these 2 families are
distantly related is under investigation.
This is possible as the abnormal gene
appears to be very rare indeed.

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Quarterly Review

CONTACT DERMATITIS XIII

The Significance of Nickel Sensitivity in Women

Nickel remains the commonest sensitizer in women. This observation was recently
confirmed in 2 large groups of patients from Scandinavia and Europe who were patch
tested to a series of common allergens. In the Scandinavian series (Magnusson et al.,
1968) 6 clinics from Norway, Denmark, Finland and Sweden patch tested 5,555 men and
women and of this whole group 5-9% were found to be positive to nickel. In the
European study (Fregert et al., 1969) 4,825 patients from Denmark, Sweden, Germany,
Holland, Italy and England were investigated and 6-7% were reported as being sensitive
to nickel. Denmark and Sweden took part in both investigations but different patients
were patch tested in the 2 groups. Although the overall incidence of nickel sensitivity
was very similar in both series there were some striking geographical differences. In
London, Wycombe, Gothenburg and Nijmegen 9-10% of all patients reacted to nickel,
whereas in Bari only 2% were positive, 4% in Munich, 5% in Copenhagen and 6% in
Lund. The low incidence in Bari may be partly explained by the fact that it was the
only centre in which more men than women were patch tested, there being nearly twice
as many men as women investigated in this clinic. Otherwise these differences remain
unexplained.

In both series nickel was reported as the commonest sensitizer in women. In the
Scandinavian group the sex ratio was recorded but no reason was given for the variation
of 15 women to one man in Helsinki compared with 2 women to one man in Bergen.

Marcussen (1959) showed that the incidence of nickel sensitivity is directly related to
the presence of the metal in the environment. In the Finsen Institute in Copenhagen,
he recorded the frequency of patients reacting to nickel, on patch testing, from 1936-
1955. During this 20 year period the overall incidence of nickel sensitivity increased
but there was a sharp drop in 1945 corresponding to the war time cessation of nickel
imports. The numbers rose again in the post war period when the metal was once again
brought into the country.

In London over the past 13 years, the incidence of nickel sensitivity has changed little
in those patients attending the patch test clinic at St John’s Hospital for Diseases of the
Skin. In 1953 in this clinic, of all the positive reactors, 27% were sensitive to nickel
(Calnan, 1956). In the European series (Fregert et al., 1969) of the total of positive
patients investigated in London, 22% were sensitive to nickel.

These figures are surprising because 95% of the 400 female patients, described by