Dr J S Pegum: I wonder if there is a bullous variant of acrodermatitis atrophicans and if so whether this case is an example of it.

Dr H Haber: Acrodermatitis atrophicans does not produce bullæ. The histology is quite different.

Dr Louis Forman: A lesion of epidermolysis which did not reach the stage of blistering would show no more than an erythematous disc. The layer of polymorphonuclear leucocytes seen beneath the epidermis could be attracted to this site by the damaged collagen.

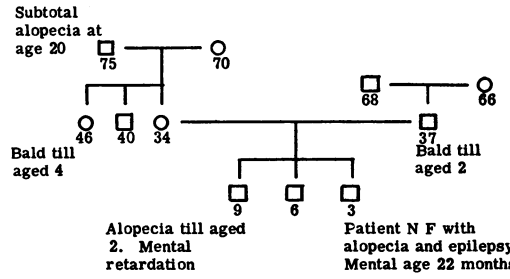

Fig 1 *Pedigree of familial congenital alopecia, epilepsy, mental retardation with unusual electroencephalograms*

Familial Congenital Alopecia, Epilepsy, Mental Retardation with Unusual Electroencephalograms

E J Moynahan FRCP

N F, male, born 4.9.58

History: Hairless at birth. Very scanty downy scalp hair and eyelashes have appeared in the last four months.

Grand mal convulsions – three with fever when six months old. Recurring since 2¾ years old every two to three weeks. Admitted in status epilepticus to St Helen's Hospital, Hastings (Dr L G Scott).

Milestones: Sat at 7 months, walked at 13 months. Little interest in surroundings and food till aged 2½. Talked first at 2½ – says few words now. Used spoon at 2¾.

No other symptoms. Playful with mother and other children. No head injury.

Past history: Measles. *Family history: See* Fig 1.

On examination: Large boy. Coarse features. Short, fine, blond scalp hair. No eyebrows. Sparse eyelashes. Skull circumference 21½ in., prominent suture lines. Fontanelles closed. Fingers stubby. C.N.S.: Retarded speech; says 'hello', 'silly' and 'two'. Behaviour friendly. Throws toys.

Investigations: Skull X-ray normal. Bone age (left wrist) 2½ years. EEG (Fig 2): Gross generalized abnormality of unusual kind – ? due to developmental anomaly. Lumbar puncture: normal pressure, protein and cells. Urine: amino acids normal; no phenylketones. Blood urea 30 mg/100 ml, alkaline phosphatase 16 K-A units, serum calcium 10·5, phosphorus 5·1 mg/100 ml.

Scalp biopsy: Small, scanty hair follicles. Some contain rim of keratin but no hair, others a small hair. No inflammation.

Treatment: Phenobarbitone gr ½ b.d., Epanutin 100 mg b.d. controls epilepsy.

The President: The extraordinary thing is that these patients grow their hair again.

Dr S Shuster: Is there any other generalized abnormality?

Dr E J Moynahan: He has been seen by several pædiatricians and there is no other defect at all. We are awaiting further biochemical results.

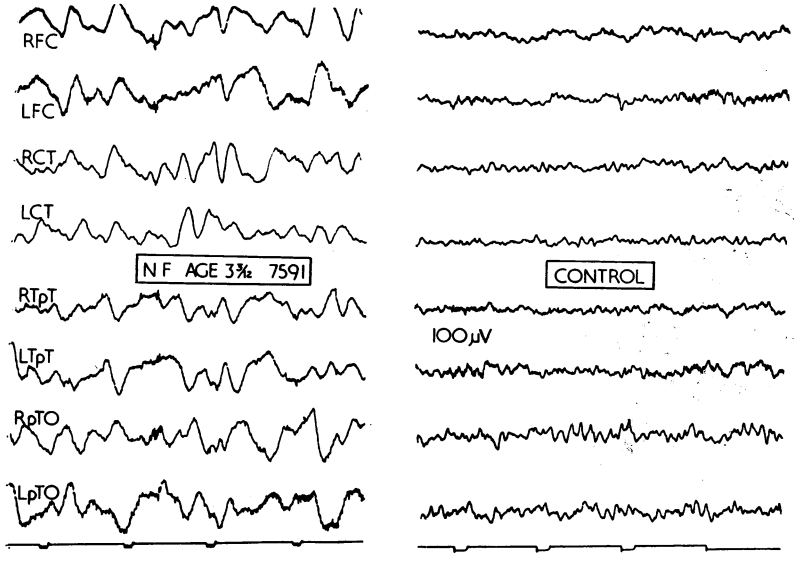

Fig 2 *EEG of N F. Note absence of α rhythm, generalized slow activity between 1 to 4 c/s mixed with a small amount of low-amplitude fast activity. No clear differences between the anterior and posterior half of the head. No spikes or complex wave forms appeared at any time. Normal control of same age for comparison*

Postscript (9.4.62): Blood lead 42 μg/100 ml; repeated examination of urinary amino acids normal.

Since the patient was discharged from Great Ormond Street it has been learnt that his mother had an epileptic seizure for the first time. It is proposed to examine the siblings and other members of the family for abnormal EEG.

The association of congenital familial alopecia, oligophrenia and epilepsy with an unusually bizarre EEG constitutes what appears to be an undescribed syndrome. The genetics suggests that the patient may be exhibiting the effects of the gene in the homozygous state, a gene which in single dose merely inhibits hair growth during late intra-uterine life and early infancy.—E J M.

Pyoderma Vegetans

M A Smith DM (for G C Wells FRCP)

H B, female, aged 68

History: Granulomatous lesion of upper lip for eighteen months slowly enlarging (Fig 1). Healthy until 1958 when she had diarrhœa, the bowels opening seven or eight times daily. Seen at the Gordon Hospital and treated with oral steroids for a few months.

1959: Developed sores, like styes, on the lower lids of both eyes. These started as small pustules which spread along the lids and became crusted and weeping. They persisted for one year, were treated with various ointments and then disappeared. A similar lesion also appeared in the right nostril and then on the upper lip.

Since 1958 bowels have opened three or four times daily, the stools formed but loose with no pain, slime or blood. She has gained 7 lb in weight over the past two years.

Investigations: 13.7.61: *Staph. pyogenes* cultured from lip lesion, sensitive to all antibiotics.

14.7.61: Hb 89%, W.B.C. 5,400 (neutros. 75%), E.S.R. 6 mm in one hour (Westergren). Biopsy (14.7.61): 'Section shows gross epithelial hyperplasia and cellular infiltration in the dermis with a few acantholytic cells'. Sigmoidoscopy: Granular red mucosa with punctate hæmorrhages to 15 cm. Barium enema (21.7.61): Extensive ulcerative colitis of the whole of the large intestine.

Recent treatment: Injection of suspension of hydrocortisone 20 mg was given into the left side of the lip lesion on 3.8.61 with some resolution for a few weeks. Hydrocortisone retention enemas 100 mg b.d. were given for six weeks in July to September 1961 and were discontinued following an attack of phlebitis in her right leg. The granuloma of the upper lip has approximately doubled in size during the last six months.

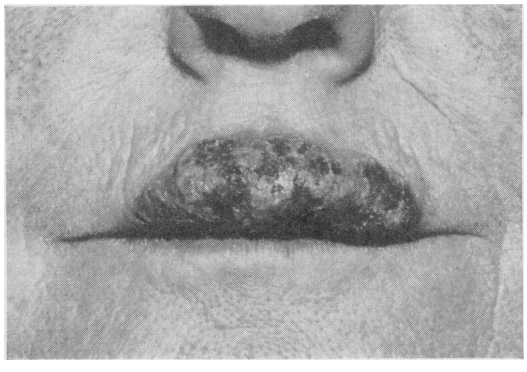

Fig 1

Dr Louis Forman: I recall a doctor aged 28 who was said to have had ulcerative colitis, the sigmoidoscope showing a vegetating mucous membrane. Subsequently he developed similar lesions in the mouth considered to be pemphigus. Scrapings showed acantholytic cells. The disease was controlled by prednisolone. Would Dr Wells consider the diagnosis of pemphigus vegetans?

Dr G C Wells: The histology in this case shows papillomatosis and dense infiltrate of polymorphs, eosinophils and plasma cells. The thickened epidermis shows micro-abscesses and acantholysis. This histology is compatible with a diagnosis of pemphigus vegetans, but I think that the same changes may be seen in pyoderma vegetans. Since this woman has ulcerative colitis I favour the diagnosis of pyoderma vegetans and I consider that the case is similar to those described by Brunsting & Underwood (1949, *Arch. Derm., Chicago*, **60**, 161).

The President: She did not have a bullous phase at all. There was a somewhat similar histology shown here last March by Dr B Woods. The same question arose as to whether we were dealing with a case of pemphigus vegetans, but I thought the evidence favoured the diagnosis of pyoderma vegetans.

The following cases were also shown:

Progressive Hæmangioma with Family History of Epistaxis
Dr K M Witham (for Dr G B Mitchell-Heggs)

Erythema Elevatum Diutinum
Professor C D Calnan

Leg Ulceration Associated with Blood Dyscrasia
Dr S Schofield & Dr John Everall (for Dr G B Mitchell-Heggs)

Tertiary Yaws
Dr D D Munro (for Dr M Feiwel)

Bilateral Gangrene of Scalp due to Giant Cell Arteritis
Dr M W Greaves (for Dr M Feiwel)

Mycosis Fungoides
Mr J P Williams (for Mr H H G Eastcott)