

NOTE ON A CASE OF PREMATURE SENILITY (PROGERIA).

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IN the present defective state of our knowledge of the factors regulating growth and development it seems worth while to make a

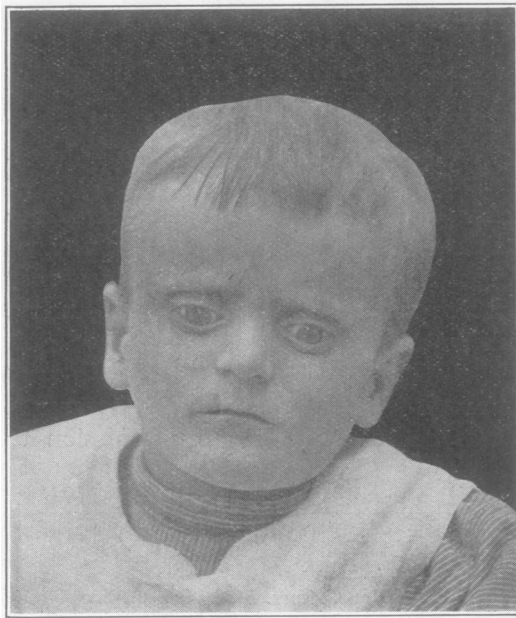


FIG. 1.—Patient at the age of four years.

(*Photograph taken by Dr. Brushfield.*)

record, however fragmentary and incomplete, of any case in which the normal mechanism is manifestly out of gear. Accordingly I offer for consideration the following brief note on a case which cannot readily be assigned to any of the recognized clinical types of disordered growth.

The patient is a little boy of 9 years, who immediately arrests one's attention by his peculiar appearance, presenting as he does an incongruous mixture of childhood and old age. In size and manner of dress he looks a child of five, but his shrunken face and deep-set eyes are those of an old man.

Family History.—This, so far as it is known, tells us nothing, though phthisis occurs among the antecedents on both sides.

Personal History.—The patient is the fifth child in a family of eight, and was born in May, 1912. The confinement was a difficult one, and his mother states that right from birth the child looked old and his appearance was peculiar.

He is said to have been thrown out of a push-cart on to his head at the age of ten weeks. Shortly after this accident he developed

some wasting disease, and was treated in different hospitals for a long period without a cure being effected.

When two years old he suffered from an ulcerated mouth, and his lower incisor teeth fell out spontaneously. He was admitted in an ill-nourished marasmic condition to the Fountain Mental Hospital in May, 1916. He was then four years old, and weighed only $28\frac{1}{2}$ lb. His case-sheet records that he was wet and dirty in his habits, and had to be fed and dressed. He was, however, able to walk, and could say a few words.

In January, 1917, he began to gain a little in weight, and to show some mental improvement also. By March, 1920, the child had improved so much that it was possible to transfer him to the Darenth Training Colony.

He has at one time or other suffered from most of the exanthemata, and it is not surprising that he has been troubled with an intermittent otorrhoea from an early age. The anterior fontanelle closed very late. He was late in walking and late in passing all the other milestones of development.

Present State.—As previously mentioned, the outstanding point about the patient is his old-mannish appearance. In stature he is stunted, his height being 41 in. The head is hydrocephalic in shape, with a

circumference of $19\frac{1}{2}$ in., as compared with the age average of $20\frac{1}{2}$ in. An *x*-ray photograph shows a normal sella turcica.

The scalp is covered with hair, but the latter is not abundant and appears to be growing thin, especially in the region of the temples. There is a small patch of white hair near the occiput.

His face as a whole is small compared with the cranium. The eyes are sunken like those of an old man, but there is no arcus senilis. The nose is diminutive, and the root of the nose is retracted. The lower jaw is relatively very poorly developed. The angle is subnormal; it would almost appear as if Meckel's cartilage had persisted

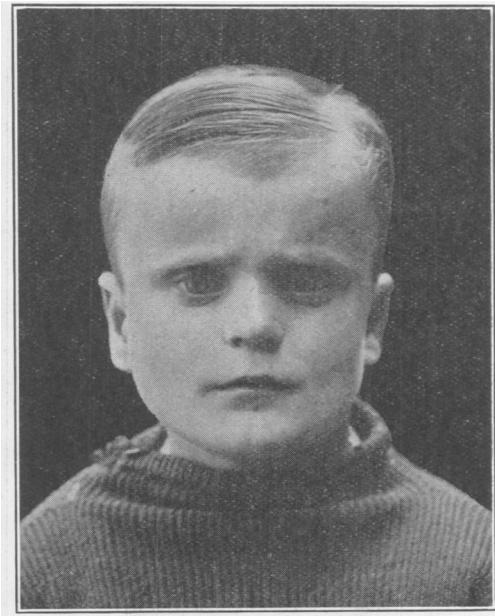


FIG. 2.—Patient at the age of nine years.

unchanged. The alveolar ridge is shallow and atrophic, analogous to the condition met with in old age. The first permanent molars and the upper central incisors are normal; the lower central and lateral incisors are absent. As a result of the defective development of the lower jaw the upper lip overhangs and overlaps the lower.

His tongue is small in size, and long and narrow in shape. The tonsils are not enlarged. His neck is distinctly small, and the skin over it is wrinkled and pigmented. The thyroid gland is not palpable.

A striking feature is the redundant and wrinkled skin covering the abdomen. The testicles are undescended. The hands and feet

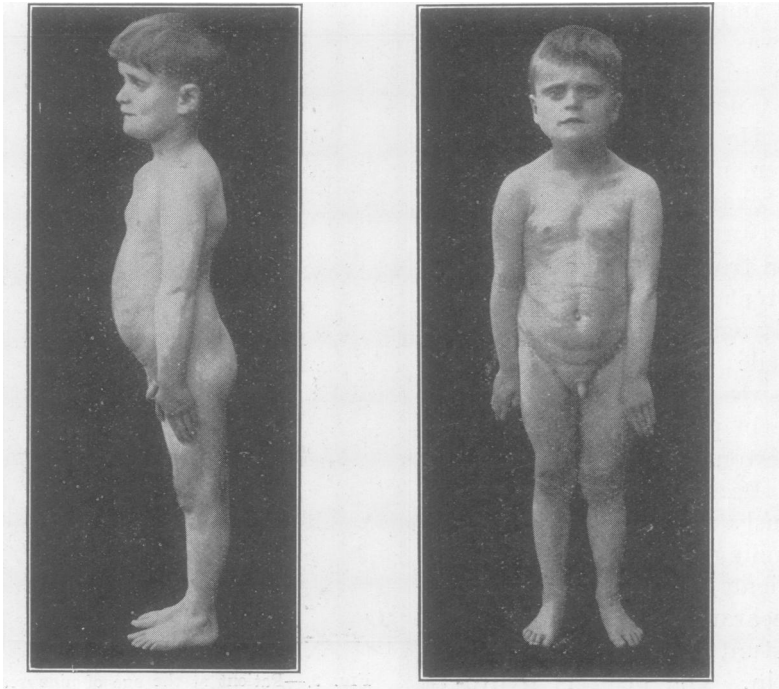


FIG. 3.—Patient at the age of nine years.

are cast in a fine mould, and are disproportionately small. Nurses have always remarked that the patient has doll's hands. His muscles are firm and well developed for his size. The recti abdominis stand out very prominently.

Neurologically there is nothing to note. The pupils are equal and react to light and accommodation. The knee-jerks are active, and the plantar reflexes are flexor. There is no sensory loss. The percussion outline of the heart is normal, and there are no signs of valvular disease. His pulse-rate is 86, with a regular rhythm. No

evidence of disease can be detected in the lungs, and the abdominal organs appear to be normal. The blood gives an absolutely negative Wassermann reaction.

In his psychical development one finds little correlation with his physical appearance of old age. Within his limitations the boy is bright and alert and takes an interest in the daily happenings in the ward. He plays with the other children, and is not specially sedate or grown-up in manner.

His mental age is 4 years and 2 months. His intelligence quotient is 46, bringing him into the imbecile class.

Discussion.—There is certainly more than enough in the patient's personal history to account for a general retardation in his development. But it is the peculiar nature and distribution of the abnormalities that is interesting, suggesting that we have to do with a disturbance of a special growth-mechanism.

It will be observed that in many respects the patient presents a picture which is the reverse of acromegaly. All those parts which are over-developed in acromegaly are found to be under-developed. The retraction of the root of the nose may be contrasted with the great boss which is thrown out from the frontal bone in a similar situation in acromegaly. In acromegaly the lower jaw is greatly hypertrophied and is thrown forwards, so that it often projects below the upper jaw. In the patient the lower jaw with its alveolar process is markedly atrophied. The circumference of the neck is distinctly small, while in acromegaly it is notably large. In acromegaly the hands and feet are above all uniformly enlarged, while in the patient they are uniformly diminished in size. An interesting question for consideration is how far the present case is identical with the condition named progeria by Hastings Gilford.¹

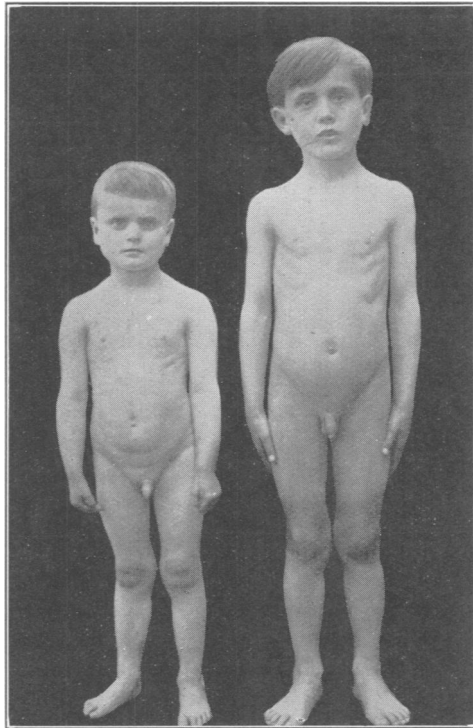


FIG. 4.—A normal boy of the same age is introduced on the right for comparison.

It seems to me that the two conditions are closely related, the essential connection between them being the fact that they both present, as it were, the obverse side of the medallion of acromegaly. In progeria we have the same wrinkled skin and the same appearance of senility as in the present case, and in both we find indications of mixed premature and immature development. There are a number of minor differences; for instance, in the case described by Hastings Gilford baldness was one of the most striking signs. Further, there was no mental deficiency, and the psychical attributes of old age appear to have been more marked.

REFERENCE.

- ¹ GILFORD, HASTINGS, "On a Condition of Mixed Premature and Immature Development", *Med.-Chir. Trans.*, lxxx, 17.



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