THE INCIDENCE OF CUTIS VERTICIS GYRATA IN THREE LOW-GRADE MENTAL DEFECTIVES

By

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THE earliest case of cutis verticis gyrata reported in the literature appears to have been that of Robert (1) in 1843. The following instances were observed by Auvert (2) in 1854 and subsequent cases were noted by Poggi (3) in 1884 and Lombroso (4) in 1890. In 1893, McDowall (5) and Cowan (6) recorded the first occurrence of this abnormal scalp development in association with a microcephalic mental defective, and since that case report several other authors such as Möller (7) in 1903 and Judassohn (8) in 1906, described similar instances. In 1907, Unna (9) presented three cases and coined the name cutis verticis gyrata. It is also known under additional terms such as "corrugated skin", "bulldog scalp" and "pachydermie occipitale vorticillée".

The cases cited in the literature were classified aetiologically by Polan and Butterworth (10) under the terminology of idiopathic and secondary types, the idiopathic types being attributed to simple hypertrophy, to developmental anomaly and to a reversion to a lower form of life, the secondary types being ascribed to pathological conditions such as inflammatory changes, naevi, tumours and various other miscellaneous diseases leading to the hypertrophy of the scalp tissues, e.g. myxoedema, cretinism, leukaemia, acromegaly, syphilis, and tuberous sclerosis. Since 1956, some three hundred cases of "bulldog skin" have been reported in the world literature.

The first case presented is that of a microcephalic, diplegic male functioning on a severely low-grade defective level (I.Q. 6) who has been at Caswell Training School for about sixteen years. He is a white patient aged 26 years in whose paternal family history a case of psychosis (grandmother) had been recorded. Otherwise no history of mental defect, epilepsy or cutis verticis gyrata was noted.

He is a poorly nourished, completely helpless, spastic, diplegic microcephalic idiot, without the power of speech and in appearance almost simian with pronounced supraorbital ridges, low, narrow receding forehead and backward-bent neck. The hair is black and of fine texture. There are eleven furrows in the scalp, antero-posteriorly arranged, averaging 3 mm. in width and 3 to 4 mm. in depth, separated by folds that range from 6 to 14 mm. in width. The folds and furrows near the midline are more pronounced than those laterally placed and they adhere to one another and are incapable of being smoothed out by traction on the scalp. In addition to these irregularities, there are others running obliquely over the higher portions of the occipital and parietal parts of the head. Over the vertex these abnormalities are most conspicuous. The whole scalp acquires a convolutional, cerebriform appearance, in which the folds and furrows of the scalp resemble the gyri and sulci of the brain surface.

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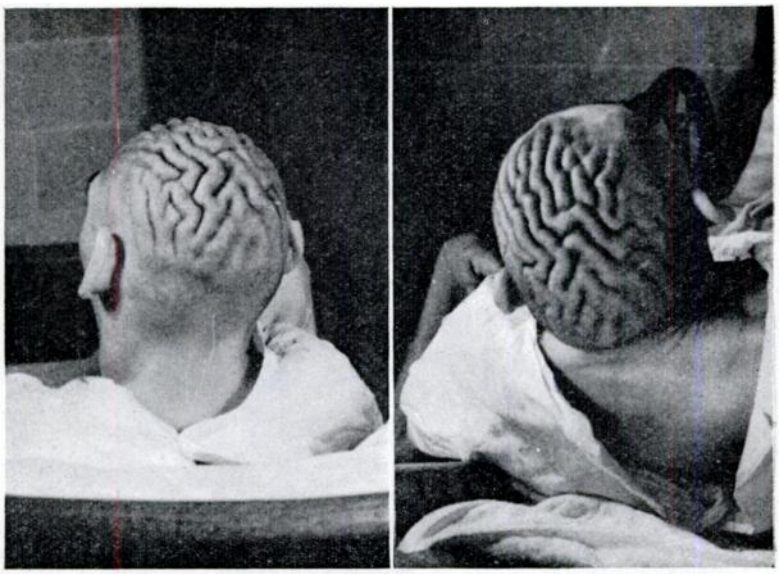
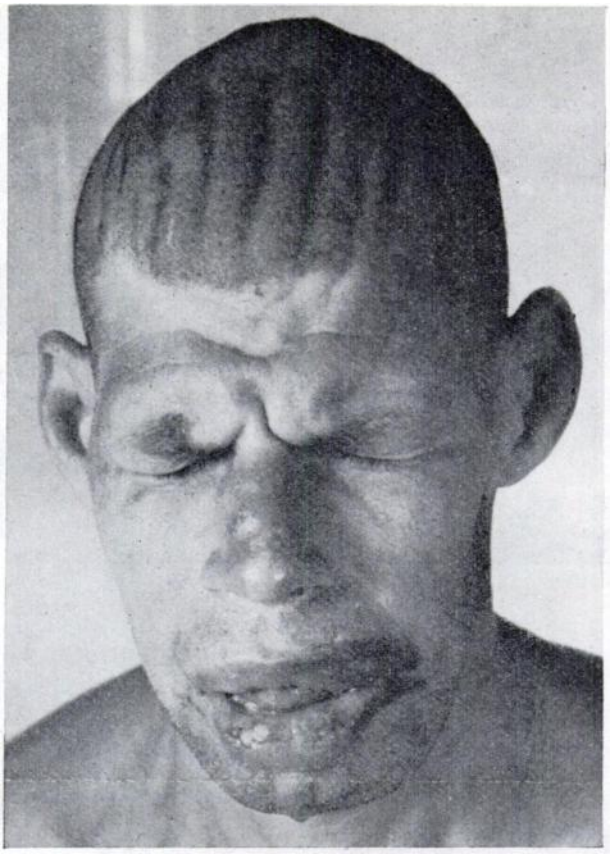
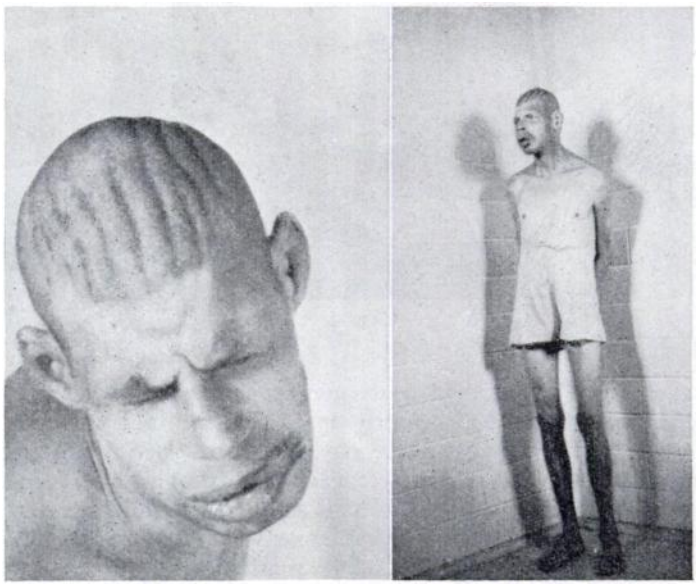


CASE I

The second case is that of a white microcephalic patient aged 23 who is severely defective (I.Q. 5). He is poorly developed, thin and of small stature. He stands about the ward most of the day, is without speech, requiring toilet and feeding care. His cranial circumference is 16 inches, his hair dark and fine and at birth he weighed 4½ pounds. There is a negative family history regarding the incidence of psychosis, mental defect, epilepsy or "bulldog skin".

Beginning near the vertex and extending anteriorly over the frontal area are nine furrows in the scalp, each about 2 mm. in depth and 2 to 3 mm. in





CASE II

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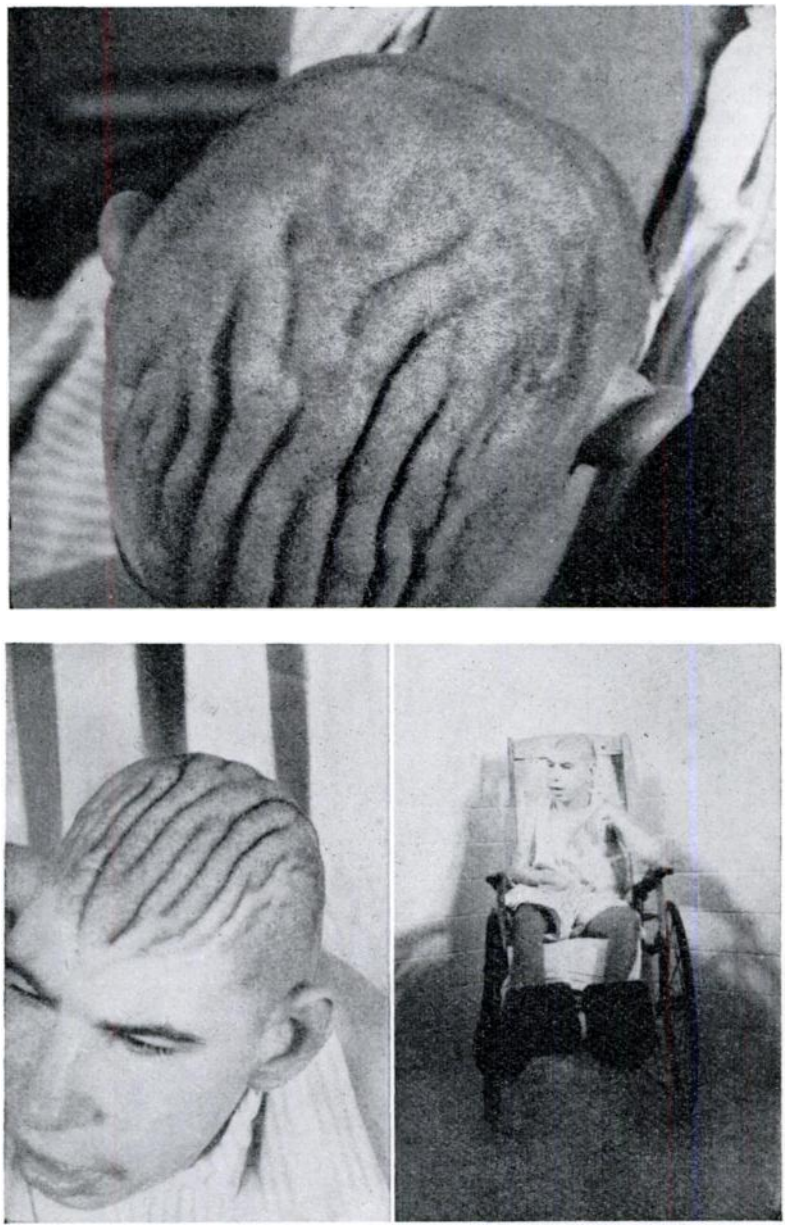
width, separated from each other by eight folds 8 to 13 mm. wide. The scalp is not movable over the underlying surface. The temporal, parietal and occipital scalp areas are not involved.

The third case is that of a white microcephalic patient aged 21. He is a spastic, diplegic cripple with multiple contractures who is severely defective (I.Q. 5) and in need of nursing care, and supervision. His cranial circumference is 15 inches, and at birth he weighed 9 pounds. At times he becomes very noisy and destructive requiring restraint and sedation. There is a negative family history.



CASE III

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Over the frontal area extending to the vertex, there are eight ribbon-like bands each varying in width from 6 to 12 mm., separated by nine furrows 2 mm. wide and 4 mm. in height, commencing almost from the anterior hairline of the forehead. The scalp over the occipital and parietal regions is normal.

Summary

Three cases of cutis verticis gyrata are described in 3 severely defective, microcephalic males, two of whom have associated spastic, diplegic complications and contractures.

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