

## **A case of microcephalic imbecility : with remarks / by G.E. Shuttleworth.**

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A CASE OF MICROCEPHALIC IMBECILITY,

WITH REMARKS.

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BY G. E. SHUTTLEWORTH, B.A., M.D., &c.,

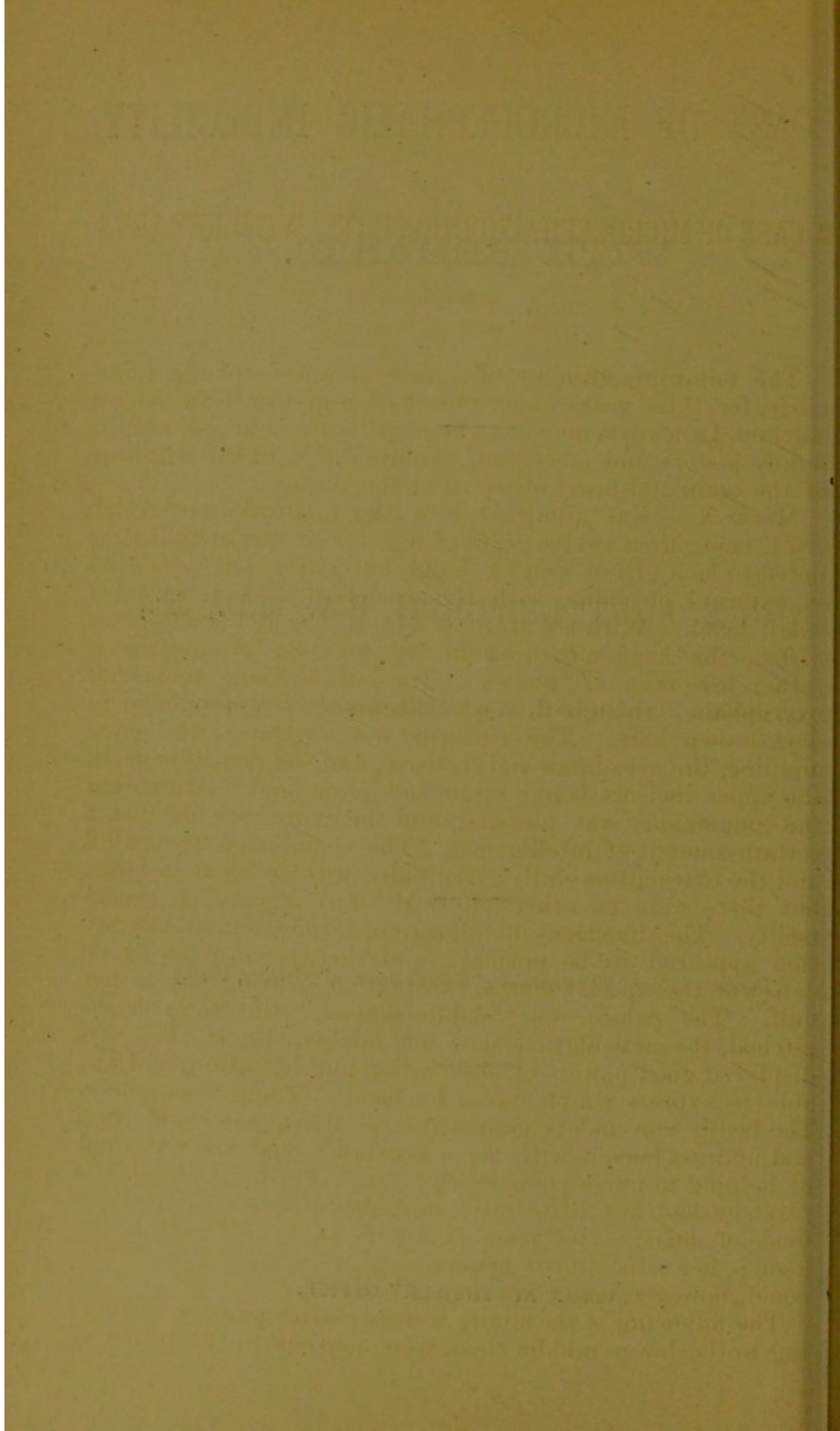
Medical Superintendent, Royal Albert Asylum, Lancaster.

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## A CASE OF MICROCEPHALIC IMBECILITY, WITH REMARKS.

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The following account of a case of microcephalic imbecility, for three years under observation in the Royal Albert Asylum, Lancaster, may be of interest, both on account of the family history and of certain abnormalities in the structure of the brain and heart observed at the autopsy.

Mary X— was admitted into the institution in April, 1874, being then twelve years of age. She was at that time noted to be a girl of slender build, but fairly well developed in external physique, with the exception of a remarkably small head. With a stature of  $61\frac{1}{4}$  inches and a weight of 87lbs., the head measured in its greatest circumference rather less than 17 inches. Its outline was somewhat oxycephalic, though this was disguised by a profusion of dark-brown hair. The features were regular, the nose aquiline, the eyes large and lustrous, and the dentition good, the upper incisors being large and prominent. Altogether the countenance was pleasing, and the expression denoted a certain amount of intelligence. The complexion was pallid and the extremities chilly; the pulse was deficient in force, but there were no other physical signs of cardiac abnormality. The functions of digestion, respiration, and excretion appeared to be normal; and there seemed to be no defect of the special senses, except that hearing was rather dull. The palate was "saddle-shaped," the voice high-pitched, the articulation slow but fairly distinct. The girl displayed good powers of observation and imitation, and was able to express simple ideas by means of short sentences. The bodily movements generally were slow; she could dress and undress herself with some assistance, and she was fond of helping to nurse younger children. For ordinary child's play she had but little taste, and she was not particularly fond of music. Her memory for persons was said to be good; her educational attainments were almost nil. She could, however, count her fingers correctly.

The following is the family history:—Her parents, belonging to the lower middle class, were persons in the prime of

life, of fine physique and apparently vigorous constitution, temperate in their habits, and thoroughly intelligent. There was no abnormality of cranial conformation in either parent. There was no account of hereditary neurosis or mental defect on either side; but the father and mother were first cousins, and the offspring of the marriage was as follows. The first-born child was a girl, the subject of the present notice; the second a boy, perfectly healthy and intelligent; the third and fourth twin boys—one healthy and intelligent, the other microcephalic and imbecile. No special cause could be assigned for the occurrence of the microcephaly, except, as the mother suggested, it might have been determined in the first case by the anxiety resulting from what she had heard as to the bad effect of marriages between cousins. No unusual incident seems to have occurred during either of the pregnancies which issued in the birth of microcephalic children.

Mary X— progressed slowly but perceptibly under institution training, improved in distinctness of articulation, and in extent of vocabulary, learned to read a few words and to form letters in a copy-book, and developed some capacity for simple domestic work. She attained considerable aptitude in calisthenic exercises. She grew fast, but did not become more robust, and her temperament remained sluggish. Her disposition was gentle, and she was affectionate towards those who had the care of her. She never menstruated. In May, 1877, she began to suffer from obscure pains in her limbs, and afterwards developed symptoms of phthisis, from which she died in August of the same year.

The autopsy, at which I had the valuable assistance of my friend, Dr. Ireland, of Larbert, disclosed the following appearances.—

The body was much emaciated; 67 inches in height; 24 inches in chest-girth at level of mammæ.

The head-measurements were as follow:—

- (a) Circumference . . . . .  $16\frac{3}{4}$  inches=425 millimetres.  
 (b) Antero-posterior—  
 (from nasal notch to occipital protuberance)  
 1. Over vertex (tape-measure)  $10\frac{1}{2}$  inches=261     ,,  
 2. Basically (calliper measure)  $5\frac{3}{4}$  inches=146     ,,  
 (c) Transverse—  
 (from tragus to tragus)  
 1. Over vertex (tape-measure) 11 inches=280     ,,  
 2. Basically (calliper measure)  $3\frac{7}{8}$  inches= 97     ,,

The circumferential contour showed some asymmetry of skull, the right side being slightly larger than the left. This asymmetry was specially noticeable at the base, the right orbital plate and the right half of the sphenoid bone being broader by quarter of an inch than the corresponding structures on the left side. The crista galli was set obliquely pointing to the left side; and the anterior and middle fossa of skull seemed slightly more capacious on the right than on the left side. The cranium varied in thickness, measuring no more than one-eighth of an inch in some portions where sawn through for the removal of the calvarium. The sutures were all closed except the coronal, and the ossification of that was well nigh complete. The arch of the palate was highly vaulted.

*Encephalon.*—There was some fulness of the superficial vessels of brain and slight sub-arachnoid effusion. The cerebrum, cerebellum, and pons together, weighed immediately on their removal from the skull  $21\frac{1}{2}$  ounces, including some little fluid. The general appearance of the encephalic mass and the arrangement of the convolutions are well shown in the accompanying wood-cuts (Figs. 1 and 2) taken from careful drawings, for which I am indebted to Dr. A. H. Young, Demonstrator of Anatomy in Owen's College, Manchester. An asymmetrical character is observable about the anterior lobes of brain corresponding to that of the skull, the right side slightly exceeding in size the left. But the most striking peculiarity is the deficiency in size of the cerebral hemispheres (see fig. 1). These are, both absolutely and relatively, abnormally small, so that when the encephalon is viewed from above, the cerebellum is seen to be uncovered over the greater portion of its upper surface. The deficiency in size of the cerebrum is mainly due to an imperfect growth of the hemispheres backwards and downwards, *i.e.*, in the occipital and temporo-sphenoidal regions respectively. As a consequence, not only is the cerebellum left exposed superiorly, but the Island of Reil also remains uncovered, whilst the two limbs of the Sylvian fissure are separated at their origin by nearly half-an-inch (see fig. 2).

Of the lobes of the brain, the frontal and parietal are comparatively well developed, and their convolutions, though coarse, are well-defined and normal in arrangement. The temporo-sphenoidal lobes are small and deficient anteriorly, and the convolutions and fissures are not so well marked as those of the frontal and parietal lobes; the normal arrange-

ment of the gyri in three antero-posterior tiers is, however, distinctly traceable. The occipital lobes are quite rudimentary, and it is impossible to detect evidence of the normal fissures and convolutions. The Island of Reil presents a flat and simple surface, the radiating gyri (*gyri operiti*) being entirely wanting.

On the inner surface of each cerebral hemisphere the corpus callosum is seen to be normal in extent. The callosomarginal fissure, however, terminates about an inch behind the genu of the corpus callosum, and is then broken up by secondary gyri. The calcarine fissure joins the internal parieto-occipital at a very acute angle, and the enclosed cuneate lobe is correspondingly small.

The cerebellum is relatively large (as compared with the Cerebrum), and is in all respects well developed. Its superior vermiform process is unusually distinct.

The Pons Varolii presents no obvious peculiarity of structure.

After being kept in spirits for twelve months, the entire encephalic mass is found to weigh only  $12\frac{3}{4}$  ounces; and of this the cerebral hemispheres weigh  $9\frac{3}{4}$  ounces; and the cerebellum, pons, &c., 3 ounces. The cerebellum alone may be estimated at  $2\frac{3}{4}$  ounces; so that its ratio to the cerebrum may be stated as 1 : 3.5, the normal ratio (according to Gray) being 1 : 8.

On section the grey matter of the convolutions appeared to be of normal proportionate thickness. My friend, Dr. Fletcher Beach, has kindly furnished me with the following note of microscopic appearances observed by him in a section from the frontal region:—

“The cortical layers are not so well defined as in the normal brain. There is a predominance of the round cells common to the second layer, and they are smaller than usual. The pyramidal corpuscles are fewer than normal, and their outline is for the most part rounded. Few have processes, and these are not well-developed. Some sections present a curious honey-comb appearance, due apparently to the degeneration and consequent non-staining of many of the large cells so commonly found in the brains of idiots of low type.”

It remains to add that the thorax and abdomen disclosed evidence of tubercular disease in the lungs, and in the glands of the mesentery. The heart weighed but six ounces; the walls were thin, but the valves competent. The foramen ovale was imperfectly closed, an aperture admitting the passage of a slender probe being found in the thin membrane

separating the auricles. The breasts were undeveloped, and the sexual characteristics generally immature.

*Remarks.*—This case is interesting from an embryological as well as a psychological point of view. The deficiency in the development of the posterior parts of the cerebral hemispheres, corresponds, in certain respects, with other recorded observations of microcephalic brains. Thus, in the remarkable case of Antonia Grandoni, a microcephal, whose head measured only 13 inches in external circumference, it is stated on the authority of Dr. Adriani, that the cerebral hemispheres, themselves 100 millimetres in length,\* “were shortened posteriorly, so that the cerebellum was left uncovered for about 70 millimetres. All the cerebral lobes were small; the parietal and occipital were smaller in proportion to the frontal and temporal lobes.” Again, in Dr. Valenti’s case of an idiot, whose cranial circumference was 13 inches 9 lines, Dr. Ireland states† “that the most striking features of the brain are the smallness of the cerebrum compared with the cerebellum, and the imperfect development of the parietal and occipital lobes, as compared with that of the frontal and temporo-sphenoidal.” In the case of Mary X., it is to be remarked that the frontal and parietal lobes were those most developed, the temporo-sphenoidal less developed, whilst the occipital lobes were quite rudimentary. This is what one would anticipate from some arresting influence having been brought into play to check the growth of the cerebral hemispheres backward and downward, probably at or a little before the sixth month of gestation. The development of detail, however, would appear to have gone on normally, so far as formative processes were completed, and consequently the convolutions of the earlier-formed portions of the hemispheres are well marked. The frontal regions are comparatively well advanced in development, and this might, perhaps, have been predicated from the manifestation of rudimentary, yet characteristically human, intellectual functions during life. It is interesting to note the aptitude for drill displayed by this girl, as showing powers of observation and imitation, and activity of the motor centres which, according to Ferrier, are found in the frontal and parietal lobes. According to the same authority, the auditory centre is placed in the superior temporo-sphenoidal convolution, and it is at least remarkable

\* Ireland, “Idiocy and Imbecility,” p. 108.

† Op. Cit., p. 118.

that, in this case, defect of hearing co-existed with an ill-developed condition of the temporo-sphenoidal lobe. The phenomena observed during life do not furnish any evidence in support of Ferrier's suggestion as to the relation of the occipital lobes to the visceral sensations, which, in this case, appear to have been normal, though the occipital lobes were rudimentary in character. The sluggish temperament noted, seems to have depended more upon feeble circulation, to which the imperfect state of the heart contributed, than upon deficiency of the nervous system. Speaking generally, microcephalic idiots, whose physical health is good, are remarkable for muscular activity; and this is well illustrated in the brother of Mary X., who is a healthy and nimble boy of 10, with a head measuring 17 inches in its greatest circumference. It may be interesting to add that his twin brother's head measures 21 inches in circumference, and that, while the stature of the imbecile boy is 47 inches, that of his more fortunate brother is 51 inches.

FIG. 1.—General view of Microcephalic Brain seen from above Actual size.  
A Parieto-occipital fissure. B Horizontal fissure. C Ascending limb of Sylvian fissure. D Fissure of Rolando.

FIG. 2.—Semi-diagrammatic view of Microcephalic Brain, showing general arrangement of convolutions (right side). F.R. Fissure of Rolando. P.O. Parieto-occipital fissure. F.S. Fissure of Sylvius: 1 Horizontal limb; 2 Ascending limb. I.R. Island of Reil.

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