Short Notes and Clinical Cases.

CONGENITAL DEFICIENCY OF CONSCIOUS AUDITORY PERCEPTION OF WORDS (WORD-DEAFNESS):

WITH REMARKS ON OTHER DEFICIENCIES OF CONSCIOUS PERCEPTION.

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THE patient, an English boy (J. B.)¹, age 3 years, is of at least average intelligence and general development, excepting for the defect which I should prefer to call 'congenital deficiency of conscious auditory perception of words,' generally known as word-deafness or 'surditas verbalis.' By analogy I should prefer to call the corresponding condition of word-blindness ('cæcitas verbalis') 'congenital deficiency of conscious visual perception of words.'²

The only words the boy can utter are 'da-da' and 'ma-ma,' but he cries and laughs in the normal way with appropriate facial expression, and expresses his satisfaction or interest or excitement by ordinary infantile sounds, which are rather musical, not loud, and never disagreeable to hear. He can call attention to or obtain what he wants by pointing and other gestures accompanied by sounds. He can certainly hear *loud noises*, for his attention can be attracted by the ringing of a bell, clapping of hands, or shouting, but he seems to react rather sluggishly, and there is no palpebral reaction to an ordinary coin-click. He does not seem particularly to like music (gramophone or 'wireless'). He is normally interested with everything he sees and from a picture-book he can pick out pictures of animals that he wishes to show to his parents. He can play ordinary games or dance with other children or with adults, is good-tempered, and never mischievous, excepting unintentionally at times owing to his great curiosity for intelligently examining everything. He apparently has occasionally said 'ta' on being given something, but ordinarily when wanted to thank anyone, only moves his lips, as if it was that he was wanted to do. He sometimes wets his bed at night, but in the day-time always makes it understood that he

¹ The case was demonstrated at the meeting of the Royal Society of Medicine, Section for the Study of Disease in Children, on January 27, 1938.

² Cf. F. P. Weber, 'Case of Congenital Word-Blindness,' *Brit. Journ. Child. Dis.*, 1917, xiv, 188.

wishes to go to the closet. When he comes home to his mother he will let her know in his own way what he has seen and experienced—for instance, using a little hammer to show that his knee-jerks have been examined at the hospital. When the baker rings the bell he will point to where the baker's book is lying, and he helps his grandmother in getting the spoons, etc., ready for tea. Although, of course, he cannot lip-read, he always somehow understands what his grandmother tells him to do—doubtless by her facial expression and gestures.

The boy is well-built, and nothing abnormal is found in his thorax, abdomen, limbs or urine. His pupils react to light, and his knee-jerks are normal. Nothing abnormal was seen by ophthalmoscopic examination (Dr. C. Markus). Dr. W. Wilson finds the mouth, fauces and ears (otoscopic examination) normal, and there is a normal response to the caloric test (cold syringing—left ear only tried). There is no history of any trauma or serious infection. His mother was in a motor-car smash when she was in the third month of pregnancy. The blood-serums of the father and mother both give negative Wassermann and Meinicke reactions. The child prefers to use his left hand, as his maternal grandmother did in childhood. He is the only child (result of the only pregnancy), as his mother was. Both father and mother enjoy good health. The father is one of a family of nineteen brothers and two sisters. There is no history of deafness, dumbness or abnormality of speech in the relatives, or of delay in learning to speak, imbecility, insanity, epilepsy, tuberculosis or any special tendency to shortness of life. There is no consanguinity between the patient's parents.

REMARKS ON OTHER DEFICIENCIES OF CONSCIOUS PERCEPTION.

In regard to the neurological and other problems connected with the subject and for an analysis of the literature, see C. Worster-Drought and I. M. Allen, this JOURNAL, 1929, IX, pp. 193, 289, and 1930, X, p. 193. These authors prefer to call the condition 'congenital auditory imperception,' and especially discuss its relation to idioglossia and other speech defects. In the present case it is of course doubtful how far the patient can consciously perceive sounds (musical, etc.) other than those connected with language. I will not enter here into all the questions of defective conscious auditory and visual perceptions apart from perception concerned with language.

In the heading I have written 'conscious auditory perception' intentionally, to show that I believe there are also *subconscious* and *unconscious*, and not only *conscious*, auditory (and, of course, other) perceptions. I admit that my terminology in this respect involves enlarging the definition of the word 'perception' so as to include what is manifested when a response of any kind follows any kind of sensory stimulus, no matter what the level in the central nervous system may be of the path between the stimulus and the resulting response or reaction. 'Perception,' in this

use of the term, may be mindless (or unconscious) as well as mental (or conscious); for convenience of terminology I postulate that perception is present (apart from any percipient mind) whenever any congenital (inherited) or acquired (conditioned) reflex action takes place.

Analogous to congenital-developmental defects in the conscious perception of words, congenital-developmental defects in the conscious perception of smell and taste probably likewise occur, but they must be even rarer, for doubtless the conscious perception of smell and the conscious perception of taste are almost as primitive as conscious perceptions of touch and pain, and are therefore extremely unlikely to be congenitally wanting or to be so weak that, like any *locus minoris resistentiæ*, they are easily lost (temporarily or permanently) from various post-natal causes. Moreover, such congenital-developmental defects in smell and taste would readily escape recognition (as partial or complete colour-blindness often does, though this is not strictly analogous).

In cases of congenital dumbness or 'congenital aphasia' of the above-described kind (congenital word-deafness) it is known that the patients subsequently learn to speak—with or without marked 'idioglossia.' Adaptation in brain defects may certainly (as is well known) be wonderful, and together with adaptation the natural post-natal development of the cerebral cortex must be taken into consideration. A question arises whether in similar cases the actual commencement of speech may ever be very sudden. One could conceive that a gradual adaptive development might silently take place in the brain to such an extent that under the influence of some shock, powerful emotion or danger, speech might suddenly spring forth from a hitherto dumb child like fully-armed Athene from the head of Zeus.

A few doubtful examples to illustrate this idea have been recorded in modern times, but by far the most interesting is one considerably over two thousand years ago, namely, the oft-repeated story narrated by Herodotus, the 'father of history.' At the battle of Pteria (B.C. 546), when Cræsus, King of Lydia, was overthrown by the Persian Cyrus, a soldier in the Persian army was about to kill Cræsus, not knowing who he was, when the latter's son, who had been dumb from birth, called out: 'Do not kill the King Cræsus.'

One may explain this story in the way suggested above, instead of telling Herodotus to his ghostly face that he was uncritical in his acceptance of tales and sagas, as an English scholar somewhat irreverently put it:

'The priests of Egypt gammoned you;

It was not very hard to do.

We will not let you gammon us,

Herodotus, you ancient cuss!'

For comparison with cases of deficiency in conscious perception of words, etc., I will here shortly refer to a case in which *all conscious perception for every kind of stimuli was permanently lost after an operation under ether*

anæsthesia, during which severe general convulsions occurred, succeeded by prolonged status convulsivus. It seemed as if the whole cerebral cortical substance concerned with conscious perception had been permanently destroyed—just as the whole hepatic parenchyma has sometimes undergone selective destruction as the result of chloroform anæsthesia¹. The patient was an English boy (J. N.) born on February 17, 1927. When I saw him in 1931 he was four years old, and his general bodily development and state of nutrition were good, but he was completely ‘mindless’ and a cerebral diplegic. Up to the age of two years he had been mentally and bodily normal and his strange condition had followed the above-mentioned operation (for appendicitis) on March 11, 1929. In 1931 I described the boy’s neurological and mental condition as follows:—

When quietly asleep with his eyes closed he seems normal. When awake and, as usual, lying on his back in bed with knees, elbows and fingers more or less flexed, he does not at first sight appear very abnormal. But all his movements, which are chiefly of his limbs and head, are automatic, not volitional, and the movements are seldom sufficient to change his position from one part of the bed to another. He never, even automatically, attempts to grasp any object, such as a key or watch, when dangled against his hands. He cannot speak, though he occasionally utters animal-like, but apparently meaningless sounds. His placid, expressionless face hardly alters at all. All emotional manifestations (smiling, laughing, crying) are completely absent. His eyes sometimes appear to follow an object close to him, and he sometimes turns his head away from a very strong light, but he certainly does not consciously see. He seems not consciously to hear any sounds. He cannot, when asleep, be awakened by any loud sound. Even when awake he does not usually respond to sounds, though when called he has occasionally been observed to move his head. Dr. William Wilson, our aural specialist, who has kindly examined the child, reports that ‘no palpebral reflex occurs to any noise stimulus.’ Dr. Wilson has made the interesting observation that in this child nystagmus occurs normally on trying the caloric test.

Anyhow, one is justified in saying that the child cannot either see or hear in the ordinary conscious way. The same may be stated in regard to sensations of touch and pain. No special tests of taste or smell have been tried, but the parents state that the child does not swallow anything that they themselves think has a markedly disagreeable taste. He certainly does not feel anything in the ordinary conscious way. His reactions seem to be only automatic. Local contact with a hot object produces a slight response. He sometimes makes a noise in apparent response to a stimulus which to most persons is unpleasant—for instance, when the external auditory meatus is syringed out with cold water in order to try the above-mentioned caloric test. He occasionally coughs, though I have not observed this myself. Sometimes he swallows better than at other times, but when he is fed with a spoon the food often remains a little in the mouth till it is passed on by a reflexly evoked swallowing movement. There is no abnormal salivation. He of course passes urine and fæces in the bed under him.

¹ See F. Parkes Weber, ‘Complete Mindlessness and Cerebral Diplegia, after Status Convulsivus associated with Ether Anæsthesia,’ *Brit. Jour. Child. Dis.*, 1931, xxviii, 14-21.

Since the above notes were written the condition of the boy (now six years old) has remained precisely the same (for this information I have to thank Dr. J. Sangster Greig, Medical Superintendent of the Forest Gate Hospital).

It seems to me that intimately associated with the question of various defects in conscious perception, of congenital-developmental nature (including word-deafness and word-blindness) or resulting from traumatism (wounds, etc.) or organic disease (ischæmic, toxæmic or destructive) of various kinds, are still larger and obscurer questions of temporary defects in conscious perception (of many kinds and various degrees) associated with the different stages of somnambulism, hypnotism, hysteria, shock, sleep, the action of various drugs, and extraneous and autogenous toxins. One may instance especially the hysterical disturbances of sensation which may be induced by psychic causes and which may be removed (though perhaps often only temporarily) by electric and other shocks. But I certainly need not discuss the various possible explanations, such as temporary disassociation, disconnection or separation of cortical nerve-cells (retraction of cell-processes, etc.).