DEVELOPMENTAL APHASIA

**Also known as Congenital Word-blindness and
sometimes referred to as Alexia or Dyslexia**

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THIS remarkable developmental failure is met with in various sensory-motor affections and may be manifested in functional failure in connection with sight, hearing, kinaesthetic performance and probably in other functional activities.

We are here concerned with developmental aphasia connected with vision, also referred to as congenital word-blindness. This disability is met with in children but does not become obvious until the child has been confronted with the problem of learning to read. The condition does not depend on the degree of general intelligence but is met with in all grades of natural ability, including occasionally individuals of the highest intelligence, which renders it all the more important that the nature of the disability should be clearly recognised. It does not depend on failure of vision due to errors in refraction or difficulty in focusing, or other causes found in the eye, though such should always receive treatment when present. This condition is a failure in visual comprehension, caused by delayed development of unilateral hemispherical dominance and failure in word-memory and relates to the psychology of vision: letters and words are seen but not recognised. Fortunately aural perception is available and eagerly used by children suffering from this visual embarrassment and proves to be of great value to them.

There is a natural tendency to recovery in all children so affected which increases progressively with the child's development and education. The condition may be mild and transitory or severe and difficult to overcome. The cases form a graded series, including all degrees of severity. In the milder cases recovery often takes place in the ordinary course of school education: in the more severe cases persistent skilled instruction, undertaken early, is essential. In the Edinburgh Primary Schools about 10 per cent. of the children were found to be affected by developmental aphasia, of whom half recovered in the ordinary course of school education, while the remainder required special treatment.

Teachers of children may fail to recognise the fundamental cause of the symptoms and this was appreciated by Hinshelwood in 1896 when he wrote: "It is a matter of the highest importance

to recognise the true nature of this difficulty in learning to read . . . otherwise these children may be harshly treated or punished for a defect for which they are in no wise responsible." The thwarting effect of this disability, especially in highly intelligent children in whom it often appears, gives rise to emotional disturbances which vary according to the temperament of the child and may produce inferiority complex, a defiant attitude to instruction, carelessness or inattention from hopelessness and sometimes fits of crying. The connection between such symptoms and their fundamental cause has frequently remained unrecognised by teachers, but is evident to the trained observer.

Developmental aphasia has doubtless always been present amongst schoolchildren, though unrecognised, and an outstanding example of this would seem to be provided by the early life-history of the famous surgeon, John Hunter. Up to the age of 17, Hunter showed very clearly that he was in every respect, apart from book-learning at school, a highly intelligent boy, possessed of character and a great desire to understand the facts and the mysteries of the natural objects of country life in the midst of which he grew up. It was only when he was confronted with learning to read, that is, to comprehend facts and ideas through the recognition of printed symbols, that he failed to progress. He was sent to a good school where his older brothers, James and William, had excelled, but John could not learn to read. All his biographers insist on this remarkable fact in John Hunter's early life. In addition it is recorded of him that he suffered from prolonged fits of crying after he was beyond the age when such evidence of emotional distress is met with in boys.

The failure to learn to read and write in the case of a boy of John Hunter's ardent and enthusiastic temperament and the knowledge that his brothers had succeeded so well at the same school must have caused him great disappointment and distress.

He was the youngest in a family of ten and lived in a cultivated and intellectual circle where all enjoyed the advantages of learning from books, but John Hunter was denied this source of knowledge for which he possessed such an insatiable appetite. Nobody could understand why he failed to learn to read and write, when he could and did learn so much through other channels, by actual contact with phenomena, by the observation of things and by hearing others talk or read. His own recorded words are: "When I was a boy I wanted to know about the clouds and the grasses, and why the leaves changed colour in the autumn: I watched the ants, bees, birds, tadpoles and caddis worms; I pestered people with questions about what nobody knew or cared anything about." The enthusiasm of his genius, however, impelled him into the new fields of

personal observation and study in which he later so greatly excelled.

Only in the light of knowledge which was not possessed until 150 years later could the true nature of John Hunter's disability have been recognised as an example of developmental aphasia.

But even in modern times these cases may remain unrecognised: for example, in the case of Ronald Hall, of which he himself has given a valuable and interesting account in the *Brit. Jl. Ophthal.*, September, 1945. He says, "I can speak from experience when I say what a terrible handicap this is for a child. Although in World War No. 1 I spent over three years at the Front, I can truthfully say that I never during those years experienced a fraction of the stark terror that I did as a boy at school during English lessons lest I should be called upon to read aloud and reveal to the whole class that I could not do so." He was taken to an oculist but the condition was evidently not recognised, as glasses were prescribed but produced no effect. He tells how he recovered spontaneously at the age of 15 and emphasizes that it was not want of intelligence on his part, as he passed first into the Civil Service as a young man.

The first account of this disability appeared in the British Medical Journal, when in 1896 Morgan^{x.1} published the case of a boy of 14 who could not learn to read, though he had no difficulty with figures: the schoolmaster said of him that if the instruction had been aural this boy would have been the smartest in the school. Words written or printed seemed to convey no impression to his mind. Morgan writes to Hinshelwood, "It was your paper—may I call it your classical paper—on acquired word-blindness and visual memory published in 'The Lancet'^{x.2} which first drew my attention to the subject, and my reason for publishing this case was that there was no reference anywhere, so far as I knew, to the possibility of the condition being congenital."

Hinshelwood's paper, to which Morgan refers, describes in detail several cases of acquired word-blindness in adults suffering from disease of the brain, of which the following is an interesting example, illustrating the parallel between this condition and congenital word-blindness, *i.e.*, developmental aphasia:—

"The patient, an inspector of schools, a highly intelligent and educated man, was familiar with four languages, English, French, Latin and Greek: he had become word-blind and could no longer read English. When, however, I put a Greek book into his hand, he was both surprised and delighted to find that he could read it correctly: it was evident then that so far as Greek was concerned there was no sign of word-blindness. With Latin there were evidences of partial word-blindnesses and with

French even more so. The word-blindness in English was not absolutely complete—there was no letter blindness and short words here and there could be picked out. Thus the word-blindness extended to only three of the four languages: Greek had entirely escaped. He also read numbers with fluency and correctness. He read musical notes as fluently and as correctly as ever."

We know that there are separate symbolisations for different languages, music, drawing and the like.

Two weeks after Morgan's paper in 1896, Hinshelwood published his first communication on Congenital Word-blindness in the form of a critical note, explaining some of the symptoms in the case reported by Dr. Morgan, in the light of his knowledge of the phenomena of acquired word-blindness, and four years later, in 1900, he gave the first analysis and explanation in detail of the symptoms met with in this condition in order "to establish the diagnosis on a scientific basis and to show that the difficulties encountered in teaching children so affected to read could be overcome by patient and persistent training."

Hinshelwood published his book on Congenital Word-blindness in January, 1917.

In 1901 Nettleship^{x,3} having expressed his indebtedness to Hinshelwood for his exposition of the subject, gave notes of five cases of great difficulty or inability to learn to read which had come under his observation and been recognised by him clinically years earlier, though not published, notes of two of which are given. The first of these cases was a boy aged 11, who was brought to him in March, 1882, because he was extremely slow in learning to read and it was supposed that his eyes might be at fault. Immense pains had been taken in trying to teach him to read and he was very anxious to learn. Nettleship was surprised to find that his difficulty was quite as great with words of No. 10 Jaeger and even No. 16 as with No. 1. It was not want of accommodation, for neither the size of the print nor the distance at which the book was held had any influence on the result. He knew some words pretty well, but unless he recognised a word at sight, he took a very long time to spell it: if, however, he was helped by having the word spelt out to him quickly he pronounced it at once. . . .

The second case, also a boy, was brought in 1882: he had great difficulty in learning to read print, though he could read music well and showed considerable facility in drawing. What puzzled his parents was that he hardly knew even short words when he spelt them: he understood what was read to him but not what he read himself. It was found in this case also that large type was not read any better than small: his manner of reading was more

suggestive of incomprehension than bad sight. After an enormous amount of patience and perseverance the boy learnt to read easily.

Nettleship finally says, "Whatever may be the anatomical cause of this condition, its importance from an educational point of view both positively and negatively is obvious. If the defect is curable, the remedy will doubtless be found in methodical and persevering instruction in reading, begun at the earliest possible age.

Although it is not intended to attempt a review of the literature on this subject, some reference must be made to the more recent work. In 1936 Professor Rönne of Copenhagen^{x4} gave an address on Congenital Word-blindness in School Children, in which he confirms Hinshelwood's experience of this disability occurring in families and refers to the case of a woman who had suffered from developmental aphasia as a child and became the mother of six children, all of whom also suffered from the same disability. He had also met with the condition in three generations of the same family. He describes his method of isolating affected children from their normal school fellows. He deplores the fact that "this quite characteristic condition is almost unmentioned in either common medical literature or in the ophthalmological."

Samuel Orton,^{x5} Iowa, U.S.A., who in 1925 had written a very interesting paper dealing mainly with Left-handedness and Mirror-reading and writing in children, published in 1937 his book, "Reading, Writing and Speech Problems in Children." This work is based upon an extensive experience in American schools and is the most detailed and authoritative account of Developmental Aphasia which has appeared.

An outline map of the lateral and mesial surfaces of the occipital region of the left hemisphere of a human brain showing the distribution of the three chief types of visual cortex. From *Reading, Writing and Speech Problems in Children*. S. T. Orton (W. N. Norton & Co., N.Y., 1937).

In describing the occipital visual centre, Orton distinguishes (1) visual perception, (2) visual recognition, (3) visual association and defines the broad outlines of the respective cerebral fields with illustrations.

His interpretation of word-blindness is an excellent extension of Head's concept of speech as "Symbolic thinking and expression." The word "Strepho-symbolia" is used by Orton to indicate a deficit in psycho-visual orientation which he was able to identify in a number of cases.^{x.5a}

Emphasizing the importance of treatment in cases of developmental aphasia, Orton says: "The reading disability cases as a group form a clear-cut example of the appearance of emotional disturbances which are purely secondary to the academic obstacle. When, however, proper treatment is not instituted or the handicap is entirely disregarded because of the 'Laissez-faire' attitude adopted by many schools, the feeling of inferiority is very apt to extend to other fields so that the child approaches every task in the expectation of failure and all of his school work may lag seriously behind." He advises, however, that "failure in learning to read with understanding must not be considered a specific disability unless it is out of harmony with the child's skill in other fields—notably the ability to learn by hearing."

The research which was initiated under the Ross Foundation in Edinburgh in 1938^{x.6} was not undertaken because the subject was new, but because it had not received attention in the Edinburgh Primary Schools (the laissez-faire attitude mentioned by Orton!). While it was found in the course of these investigations that nearly 10 per cent. of children in the Primary Schools showed evidence of this disability, only about half that number required special tuition, the others recovering under the usual school training.

Miss M. Macmeeken, M.A., B.Ed., Ph.D.,^{x.7} who carried out the research for the Ross Foundation, has as a psychologist been very successful in the education of children suffering from developmental aphasia: she considers the undertaking one for a specialist only: the willing co-operation of the child must be secured and subjects selected with a view to maintaining the lively interest of the pupil who takes part in the work with the teacher alternately in writing and reading. Not only is good method necessary but an extended period of time is required to secure the result. Three school terms are usually found necessary.

In considering the underlying causes of this disability it would seem to fall into two parts which, while they react upon each other, are distinct:—

(a) Mirror reading and writing, twisting of syllables and confusion of certain letters and palindromic words, relating to

insufficiently developed dominance in one of the cerebral hemispheres: these earlier manifestations (as above noted) frequently disappear in the first few years of school life.

(b) Failure in word memory, which tends to be a more permanent and stubborn difficulty to be overcome: this may relate to the delayed development of myelination of cortical neurons, with associated delayed function.

A reference to this subject of myelination occurs in a paper which appeared in the Transactions of the Royal Society of Edinburgh in 1913^{x.8} by the late Alexander Bruce, M.D., LL.D., F.R.C.P.E., and James W. Dawson, M.D., from which I quote: "If it is true that every cell differentiates in view of a function, it is necessary to remember that it is the functioning which determines and perfects the cell differentiation. The nerve paths in the embryo remain as embryonic nerves till the function of the tract is called into play: influences which accelerate or retard the period at which nerve fibres are brought into functional activity have also an effect in determining the date of complete axial fibril and myelin differentiation. Margulies^{x.9} has pointed out that in the newly-born kitten, if the eyelids on one side are carefully opened, the optic nerve on that side myelinates before that of the opposite side excluded from the light, and numerous other instances might be given where the completion of differentiation is related to the completion of function. The fibres in the distal end of a non-united nerve remain for a very considerable time as embryonic nerve fibres, but when secondary suture is carried out they very rapidly effect a complete differentiation—in a period of time in which it would have been impossible for axis-cylinders to grow out from the central to the distal end. The differentiation proceeds, therefore, *pari-passu* with the functioning which is its determining cause. Ballance and Stewart^{x.10} think that some stimulus, afforded by the conducting impulses, is necessary in order to admit of the full development of the nerve fibres."

I wish here to express my indebtedness to A. Ninian Bruce, M.D., F.R.C.P.Ed., Lecturer on Physiological Neurology, University, Edinburgh, for directing my attention to the above-noted research in which he took part with his father.

In an address entitled "Some Recent Advances in the Study of the Brain as the Implement of Mind,"^{x.11} Professor R. J. A. Berry, M.D., F.R.C.S., discussed myelination in the normal and the abnormal brain. This prompted me to write to Professor Berry and to look up his former work on this subject. From his book, "Brain and Mind," 1928, the following passage seemed to have a special bearing on the possible significance of myelination in developmental aphasia: "Flechsig has shown that myelination

does not occur simultaneously in all parts of the nervous system, but is later in proportion as the nerve fibre (axon of the neuron) is more recent in the phylogenetic history of the animal. Not only are the entering receptor neurons of the spinal cord more numerous than the outgoing effector neurons, but they develop or myelinate earlier, in order that they may function earlier. Generally speaking, the same holds good for the neo-pallial cerebral cortex, and the last of the cortical neurons to myelinate are those associating or linking together the association areas of the cortex, and these are precisely the areas which are universally regarded as being the seats of the higher mental processes. But if these same areas are not stimulated by suitable incoming receptor impulses, their association neurons will not myelinate and consequently the areas remain throughout life veritable "silent" areas, to the detriment of the mentality of the individual.

It consequently follows, and it is a strikingly significant fact, that the general order of acquisition of the nerve properties essential to life are:—

1. Transmission of entero-ceptive impulses from viscera by means of both non-medullated and medullated fibres through the autonomic nervous system.
2. Transmission of proprio-ceptive impulses, concerned in locomotion.
3. Transmission of extero-ceptive impulses such as touch, pressure, sight hearing, etc.
4. The last series of neurons to myelinate, that is to function, are those of the cortex which correlate and control the incoming extero-ceptive impulses, and extend them through the association areas, thus making possible education, speech thought and reason. If the neurons of this series fail to myelinate, there must follow an impairment of intelligent action and a reaction to the environment on a lower plane.

The period of time occupied by the myelination of these four great developmental series is a long one, but differs very considerably in different individuals. It commences before birth and goes on, in the educated classes, until well on into middle life, because education, that is, the constant submission of the cortical association areas to appropriate incoming receptor impulses, is a powerful stimulus to the internuncial association cortical neurons. In the non-educated classes the process of myelination of these neurons ceases much earlier in life, and as a consequence they do not attain the same degree of intellectuality.

The importance of myelination is also borne out by pathology for it is a significant fact that in *multiple* or *insular sclerosis* of the

brain and cord, the axis cylinders of the areas affected remain intact whilst the myelin sheaths are destroyed. The disturbances of co-ordination accompanying this condition may, therefore, be an expression of a loss of insulated conduction.

In considering myelination in relation to the differentiated cortical visual fields, Orton refers to Flechsig's work as follows :

"Flechsig found that maturation proceeds in three distinct waves covering, in separate stages, those areas of the brain cortex whose destruction leads to the three syndromes of cortical blindness, mind blindness and word blindness and the comparable conditions in audition. Flechsig demonstrated that at the time of birth only the "arrival platforms" or first level cortices have received their myelin, that a second period of myelinization follows during the first two or three months after birth which results in the ripening of the second zone of cortex lying near by each arrival platform and that only during the final or third wave does maturation occur in the areas of the third level."

He continues: "I have emphasized this dissection of the cerebral functions into steps because it offers us some understanding of how a selective loss of reading, for example, in the adult, or a selective retardation in learning to read in a child, may occur with full visual competence in other regards."

Developmental aphasia is an isolated disability. The diagnosis is difficult and uncertain in mentally defective children, but clearly recognisable in children otherwise normally developed in relation to age. The characteristic mental defects are related to difficulties in symbolisation and in word-memory: all other faculties of recognition and recollection are normal.

All expert and experienced teachers of such children are agreed that encouragement, patience, perseverance and time are necessary and in advancement the pupil must make the pace. In all this the most important element in securing progress lies in oft-repeated practice (in reading and writing) by performance on the part of the pupil. This activity on the part of the child fulfils the requirements of cerebral stimulation referred to by Bruce and Berry in relation to the development of function.

I suggest that the localized delay or failure in the development of myelination of internuncial nerve fibres in the cerebral cortex is the most probable explanation of this developmental failure in learning to read.

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HISTOLOGICAL FINDINGS IN A CASE OF ANGIOID STREAKS

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FOR the privilege of making this report I am indebted to the late Dr. Grady E. Clay. It deals with the microscopic examination of one eye removed after death from a patient with angioid streaks in both eyes. The eye was removed under the direction of Dr. Clay and sent by him to Dr. Algernon Reese for sectioning. A complete set of celloidin sections was prepared and stained under Dr. Reese's supervision and then sent to me at the request of Dr. Clay. The hospital at which the patient died was the Grady Memorial Hospital of Atlanta, Georgia, and to the authorities of this institution I am indebted for complete copies of the hospital records including the autopsy report on this patient. The clinical record was signed by Guh H. Adams, M.D. The autopsy was performed by Abner Golden, M.D., and the report signed by Walter H. Sheldon, M.D., pathologist. For the purposes of this paper the following summary of these hospital records will suffice:

The patient, a 50-year-old coloured female was admitted to the Grady Memorial Hospital, Atlanta, Ga., Jan. 28, 1944. She was known to have had syphilis which was inadequately treated. However, the Kahn test was negative at the final admission to the hospital although in 1935 the Wassermann test was 4 plus. She was known to have had hypertension since 1935. In 1939 the blood pressure was recorded as 200/110 and at this time she began to experience intermittent oedema of the ankles. There were not marked symptoms, however, until three