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BRAIN:

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*An Atrophy of the Brain
By Fletcher Beach M.B., M.R.C.P.
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ON ATROPHY OF THE BRAIN IN IMBECILES.

BY FLETCHER BEACH, M.B., M.R.C.P.,

Medical Superintendent of the Darenth Asylum.

MANY writers speak of two forms of atrophy of the brain, and it is a convenient method of describing the disease. In the first, there is incomplete development; in the second, there is loss of nervous elements which had previously been present. Taking first that variety in which there is incomplete development, and excluding that form of it in which the hemispheres are completely wanting and vegetative life merely exists, I would remark that cases illustrating this condition are commonly found in Asylums for Imbeciles. Microcephalic imbeciles are instances of this class. Microcephaly may be general or partial. Some portions of the brain may be too small or altogether absent. I have found the convolutions of certain parts very much reduced in size from non-development, while the remaining portions are too large or normal. The occipital lobes are usually arrested in growth in microcephalic cases, and the island of Reil may be left uncovered. The corpus callosum is sometimes shortened posteriorly, and cases are on record where this portion of the brain has been wanting and the commissures deficient.

As an instance of the arrest of growth of the occipital lobes, I would refer to a case related by Dr. Shuttleworth in the 'Journal of Mental Science for 1878,' in which the frontal and parietal lobes were comparatively well-developed, but the occipital lobes were quite rudimentary.

Usually, however, microcephaly is general, and the deficiency consists in the smallness of the hemispheres. In the 'Transactions of the International Medical Congress' for 1881 I have related two cases, in one of which the brain when removed from

the body weighed only 7 ounces, yet a minute examination of it showed that nearly all the convolutions were present, though very small in size. In the other, the brain weighed $20\frac{1}{2}$ ounces. Two brains of microcephalic cases have since come under my notice, and these weighed $20\frac{1}{4}$ and $24\frac{1}{2}$ ounces respectively. The nerves of special sense are usually well-developed in microcephaly, and the ganglia of the base and the spinal cord are of nearly normal size. The cerebellum is relatively much larger than in the normal brain, often being in the proportion of 1 to 3 or 4, the ordinary relation being 1 to 8.

The following are illustrative cases :

G. L., aged 13 years, admitted into Darenth Asylum, January 19th, 1878. The father is a very hard drinker and often ill-uses the mother. He has an impediment in his speech, and so have all his side of the family. The mother is a temperate woman, but has a brother in Colney Hatch Asylum. Intemperance and hereditary predisposition to insanity are therefore present. The parents are not connected by consanguinity. G. L. had a small head when born. He did not walk till 5 years old, and has never spoken. The mother ascribes his condition to ill-usage by the father during pregnancy. He is a twin; the other child is dead, but his head was of normal size. There have been twelve children, of whom one, a girl aged 8 years, is also microcephalic. Seven have died, and of these one was microcephalic and died in convulsions.

On admission, G. L. was found to be rather thin, with a muddy complexion and a bird-like aspect. His head measured $17\frac{3}{8}$ inches in circumference (3 to 4 inches below the normal size), $10\frac{7}{8}$ inches transversely ($3\frac{7}{8}$ inches with calipers), and 11 inches antero-posteriorly (6 inches with calipers). Since his residence in the Asylum his head has increased a little in size, being now $18\frac{6}{8}$ inches in circumference, 11 inches in the transverse diameter, and $11\frac{1}{8}$ inches in the longitudinal direction; but even now the outline of it, when compared with one of normal size, shows the great difference at once. His forehead was $3\frac{5}{8}$ inches in breadth. He had good use of his limbs and was not subject to fits. In temper he was some-

what stubborn, but was easily managed. He could not speak, but could only make a sound like "ah." He was able to take his food without assistance, and could help in the ward, but he could not wash or dress himself without assistance. He had some power of observation, imitation, and attention, though but little memory. He went to school in the Asylum daily, and at the end of one year had learnt some letters of the manual alphabet, and could hold up one or two fingers to correspond with the objects shown to him. Two years afterwards he was able to read some words with the manual alphabet, could spell "cap," write "a, o, d" from memory, hold up fingers as far as four to correspond with objects, and could match colours and forms. He had also learnt to make himself very useful in household work, and could sew on buttons, and do simple repairs of clothing. Considering the size of his brain, I think we may say he has made fair improvement, though not so much as his sister, who has a smaller head.

A. P., admitted August 15th, 1877, was three years old on admission. Her parents are healthy, temperate, and not connected by consanguinity. A brother of the mother is subject to fits. The mother attributes the patient's condition to a fright which she had when four or five months advanced in pregnancy. This is the fifth child. The others are all healthy and have no mental defect. The child's head was small when born, and the mother thinks it has not increased much in size since, though the body has grown normally. She has never noticed passing objects, and has never been able to stand or hold anything. She is not subject to fits. Her thumbs have been drawn into the palms of her hands since birth.

A. P. when admitted was a fairly nourished child, of fair complexion, 30 inches in height. Her head measured $15\frac{1}{4}$ in. in circumference (5 to 6 inches below the normal size), $9\frac{1}{2}$ in. transversely, and $10\frac{1}{2}$ in. antero-posteriorly. As in the preceding case, the head has grown in size since admission, the corresponding measurements to those above now being 16, $9\frac{5}{8}$ and $10\frac{5}{8}$ inches. Her forehead is small and receding, tending to the oval in shape. In height it is 2 inches, and in width 3 inches. Her arms and legs are of normal size, the

former measuring 11 inches, and the latter 13 inches in length; she cannot stand, though she can kick her legs about. She is subject to spasmodic retraction of the limbs, and though she can move her arms, she cannot hold anything. Her forearms are contracted on the arms and the fingers are clenched. Her legs are also contracted on her thighs, and cannot be straightened. She is placid in disposition and cheerful. Her habits are faulty. Her mental capacity is of course very small, but there is some power of observation and a little of attention. She has now been $4\frac{1}{2}$ years in the Asylum, and during the whole of the time has been kept in the Infirmary, where she sits by day coiled up in a chair, and requiring to be fed, clothed, and generally attended to. She now seems to recognise her nurse, and laughs when pleased, but the improvement made is not great.

Both of these cases, and especially the latter, have heads much below the normal size, and correspondingly the brains are deficient in development. It is not, however, only the smallness of the quantity of the brain, but deficiency in quality which is involved. If we look at sections exhibited under the microscope, we shall see the great difference between a normal and a microcephalic brain. In the latter we notice the deficiency of cell processes, and the retraction of the protoplasm in the cell, accounting for the spaces so often visible. As to the causation of microcephalic imbecility, it has been held that it is due to the sutures of the skull closing in prematurely, and so preventing the growth of the brain; but, in opposition to this theory, there is the fact that many cases have been collected with open sutures. "Even in those cases where the sutures have closed in before birth, the question still remains whether the brain ceased to grow because the sutures are closed, or whether the sutures closed in because the brain ceased to grow; or lastly, whether both the brain and its coverings ceased to grow from a common cause."¹

The second form of atrophy, viz., that in which there is loss of nervous elements which had previously existed, may present itself in various forms, but the most interesting is perhaps that in which there is atrophy of one side of the brain, usually the

¹ Ireland on Idiocy and Imbecility.

left, with coexistent atrophy of the limbs of the opposite side of the body. Now, though imbecility is not necessarily the result (*vide* cases quoted by Van der Kolk), yet examples of this disease are of fairly frequent occurrence in Asylums for Imbeciles. According to this author, everything depends upon the more or less healthy state of one hemisphere of the brain. "If, as from the nature of the case seldom occurs, the inflammation and affection of the pia mater has not extended to this hemisphere, if the grey matter under the cerebral convolutions has here continued perfectly sound, there is no reason why this remaining hemisphere should not be able to act without impediment in the exercise of those functions which are necessary to our mental powers, just as one eye sees as sharply though the other be lost. But where grey matter is injured in both hemispheres, particularly anteriorly, disturbance of the intellectual faculties will be inevitable."

An inflammation of the brain, meninges, or skull during foetal life or early childhood, no doubt will cause the disease. The paralytic form of imbecility, for instance, seems to depend upon an atrophy of the brain caused by chronic meningitis or inflammatory processes in the cortical substance. Either of these causes may of course occur before birth or come on afterwards.

In the following case I believe atrophy of the brain to exist, and I think there is no doubt that it was produced before birth.

W. B., aged 9 years, was admitted into Darenth Asylum, November 14th, 1877. The parents were temperate and healthy people, and not connected by consanguinity. When the child was born, his right side was found to be powerless, in consequence, as is supposed, of the mother during pregnancy falling down and striking her side with great violence against a wall. On admission, his head was found to be flattened on the left side, but of fair size, being $19\frac{7}{8}$ inches in circumference. He limped with his right foot in walking, and the fingers of his right hand were flexed in the palm. His mental capacity was fair. He could read a few letters, print with his left hand "a" and "o," count to 20, and knew a few colours. He has made

good progress in school, so much so, that he is now able to read well from the 1st Standard, write and transcribe (with his left hand) in a book, work easy addition, subtraction and multiplication sums, and recognise all the colours and shades, and easy and difficult forms. He has progressed equally well in the tailor's shop, and, as a result of his industrial work, he is now able to straighten the fingers of his right hand, and make use of this hand in dressing himself and in his work.

The progress in such cases is nearly always fair mentally, though but little physically. In the one just quoted, greater physical improvement than usual has been made, but there are even now marked differences in length and size between the limbs of the healthy and those of the affected side.

RIGHT ARM (AFFECTED SIDE).

	Inches.
From point of acromion process to external condyle of humerus .	10 $\frac{1}{2}$
From external condyle of humerus to styloid process of radius .	8 $\frac{1}{8}$
From centre of wrist to end of middle finger	6 $\frac{5}{8}$
Circumference round centre of deltoid	8
" " " " " biceps	7 $\frac{1}{2}$
" " " forearm 1 $\frac{1}{2}$ in. in front of inner bend of elbow .	7 $\frac{1}{2}$
" " " forearm 1 in. in front of wrist	5 $\frac{6}{8}$

LEFT ARM (UNAFFECTED SIDE).

From point of acromion process to external condyle of humerus .	11 $\frac{1}{4}$
From external condyle of humerus to styloid process of radius .	8 $\frac{3}{8}$
From centre of wrist to end of middle finger	7 $\frac{3}{8}$
Circumference round centre of deltoid	9 $\frac{1}{2}$
" " " " " biceps	9
" " " forearm 1 $\frac{1}{2}$ in. in front of inner bend of elbow .	8 $\frac{3}{4}$
" " " forearm 1 in. in front of wrist	6 $\frac{1}{2}$

Every measurement of the affected arm is seen to be less than that of the unaffected one, and so with the leg.

RIGHT LEG (AFFECTED SIDE).

	Inches.
From inferior spinous process to external condyle of femur .	15
From external condyle of femur to external malleolus . . .	14
Length of sole of foot	9 $\frac{1}{2}$
Circumference of thigh 10 in. below inferior spinous process .	14 $\frac{1}{2}$
" " " calf 5 in. below external condyle	10 $\frac{5}{8}$

LEFT LEG (UNAFFECTED SIDE).

	Inches.
From inferior spinous process to external condyle of femur	16 $\frac{1}{4}$
From external condyle of femur to external malleolus	14 $\frac{1}{2}$
Length of sole of foot	10
Circumference of thigh 10 in. below inferior spinous process	15 $\frac{3}{8}$
" " " calf 5 in. below external condyle	11 $\frac{1}{4}$

In the next case the date of the onset of the paralysis is doubtful. E. H., aged 18 years, was admitted into the Clapton Asylum, May 3rd, 1875, having been transferred from the Hampstead Asylum. I am indebted to Dr. Orange, Medical Superintendent of the Broadmoor Criminal Lunatic Asylum, for the following history. The father of E. H. had died in 1868, of cancer of the bladder. The mother was a patient in the Broadmoor Asylum, having killed one of her children while insane. The parents had been temperate people and were not connected by consanguinity. There was no family history of epilepsy or paralysis. This was the eldest child. There had been one other, who was killed by the mother, as mentioned above. E. H. was of sound mind at birth, and was doing well at Southall School, but was sent to Hampstead Asylum in consequence of epileptic fits, from which she suffered. These commenced at the age of 2 $\frac{1}{2}$ years (cause unknown). Her intellect had become affected by the fits.

On admission, she was a fairly nourished girl of dark complexion, with loss of power of the right upper extremity (the forearm being contracted on the arm, the wrist on the forearm, and the fingers flexed in the palm of the hand), and weakness of the lower one on the same side. There was no means of ascertaining the cause of the paralysis, or when she was first afflicted with it. She was still having epileptic fits. She talked and answered questions with considerable intelligence, was of a quiet disposition, had pleasing manners and was a general favourite. Her mental capacity was very fair for an imbecile. She could read from the 3rd Standard, and write very well with her left hand. She had made considerable advance in arithmetic, being able to do simple and compound reduction sums. She was fond of music, and knew all the colours. From this it will be seen that she was an imbecile

of a very high type. She made progress at school in the intervals between the fits, and was able to sing at the entertainments. In October of the same year she was noticed to be getting very thin, and on examination the physical signs of phthisis were discovered. She now had attacks of petit-mal very frequently and became very emaciated. She died on the 21st of December, 1875, having been unconscious the preceding day for 6 hours. The post-mortem examination was made 12 hours after death. The calvaria was removed, and the convex surface of the brain examined in situ. On stripping off the dura mater, the left hemisphere was seen to be much wasted, and the arachnoid membrane over its surface was in places thickened and opaque. The wasting was chiefly noticeable in the frontal and parietal regions. The texture of the hemisphere was evidently altered, for, to the touch, the left was hard and firm, while the right was elastic and apparently normal. The left middle lobe was much reduced in size, measuring only 1 inch transversely, the right measuring $2\frac{1}{4}$ inches in the same direction. There did not appear to be excess of fluid in the left subarachnoid space, but as some drained away while removing the calvaria, the defect may be thus accounted for. The brain was then removed and found to weigh $28\frac{3}{4}$ ounces. A quantity of fluid drained away during the process of extraction. A depression as large as a small pear could now be seen on the convex surface of the posterior half of the left hemisphere. On slicing through the brain transversely, the left lateral ventricle was found to be enormously dilated, measuring $4\frac{1}{4}$ inches in length and 2 inches in breadth at its posterior (most dilated) part, the right ventricle measuring only $3\frac{3}{4}$ inches, and 1 inch in the corresponding directions. The depression above mentioned was now seen to be due to the roof of the left ventricle falling inwards. Its upper wall in this situation was not more than $\frac{1}{4}$ inch in thickness, the roof of the right measuring $\frac{3}{4}$ inch. In consequence of the dilatation of the left ventricle, the left choroid plexus in its course outwards dipped downwards one inch anterior in position to the right. Comparing the two hemispheres, it was found that the right measured 7 inches in length, and $2\frac{1}{2}$ inches in breadth, but the left only $6\frac{1}{4}$

inches longitudinally and $2\frac{1}{4}$ inches transversely. The right hemisphere weighed $15\frac{1}{4}$ ounces, the left only $5\frac{3}{4}$ ounces. The convolutions on the right side were exceedingly coarse, while those on the left side were smaller than usual for a child of her age. The white matter of the anterior and middle lobes on this side was reduced to a mere line. Unfortunately I did not examine the corpus striatum and optic thalamus of the two sides.

Examining now the cranium, the left side was found to be much thicker than the right, and the internal surface presented marked differences on the two sides. Thus the left anterior fossa was more prominent than the right, the left anterior lobe of the brain being correspondingly depressed inferiorly. The left middle fossa was much smaller than that on the opposite side and was pear-shaped, while the right was more quadrangular. On measuring the two, the left was found to be $1\frac{1}{2}$ inch in length and $1\frac{1}{4}$ inch in breadth, the right measuring 2 inches and $1\frac{3}{4}$ inch in the same directions. The left posterior fossa was shallow. The left cerebella fossa was larger than the right. The lungs presented cavities in the apices, and patches of tubercle and caseous material were scattered through the substance of both.

The cerebellum was not specially examined, but from the fact that the left cerebellar fossa was larger than the right, there is no doubt that the left lobe of the cerebellum was the larger of the two.

The third case, A. H., æt. 16 years, was admitted into the Clapton Asylum, May 5th, 1875, and died November 12th, 1875. Her father had died at the age of 33, of typhoid fever; the mother was alive and healthy. There was no family history of neuroses, but the aunt on the mother's side had died of phthisis. Both parents had been temperate, and were not connected by consanguinity. There were three other children who were physically weak, but who were unaffected mentally. The patient is said to have been mentally sound when born, and nothing was specially noticed (except some spots on the body, when the child was a month old, due to the father having had syphilis before her birth) until four years of age when the child had a heavy fall, and the right side of her

body became paralysed. Her intellect, however, was not noticeably affected until the age of 12 years, when she became subject to fits (cause unknown). These have continued ever since, but are slight in character. Since the onset of the fits she has become dull. On admission she was found to be emaciated and the right side of the body was paralysed. She could speak indistinctly, was quiet in disposition, but dirty in her habits. Her mental capacity was small. She could only read words of two letters, spell A B C, add to 6, count to 40, and recognise two colours. She was unable to dress, or take her meals without assistance. She went to school daily, but made little progress. She was very feeble, and frequently had attacks of diarrhoea and tonic contractions of the right side, no doubt epileptic in character. Later on, twitchings of the lip were noticed, and the left side of the body sometimes became affected. She died exhausted, November 12th, 1875. At the autopsy, the body was found emaciated and the right arm flexed on the forearm. The cranium was much thicker than normal. (I find I have not noticed whether it was thicker on one side than the other.) The brain weighed $29\frac{1}{2}$ ounces, and was smaller than normal for a child of her age. The convolutions were flattened, and the sulci diminished in depth. There was a large excess of subarachnoid fluid, especially over the left hemisphere of the brain. The arachnoid membrane was opaque in places, especially over the site of the subjacent vessels. On slicing through the brain, the lateral ventricles and the fourth were seen to be much dilated with fluid, though some had drained away, the left lateral ventricle being apparently the fuller of the two, so that the septum lucidum was a little to the right of the middle line. On comparing the two sides of the brain, the left hemisphere appeared smaller than the right, especially anteriorly. The corpora striata, optic thalami and cerebellum appeared normal and equal in size, but this may have been due to the atrophy of the hemisphere being small in amount.

From the cases just given, as well as those related by Van der Kolk, Dr. Taylor in Guy's Hospital Reports, and others, it appears that the usual appearances found post mortem are: thickness of the cranium, opacity and thickness of the mem-

branes, effusion of serum into the subarachnoid space, sometimes into the ventricles and atrophy of one hemisphere, including the corpus striatum, optic thalamus and pons of the affected side. Since the fibres of the superior peduncles of the cerebellum undergo a complete decussation beneath the upper pair of the corpora quadrigemina, and those of the middle peduncles decussate in the pons varolii, while the fibres of the pyramids of the medulla have their well-known crossed direction, there is atrophy of the cerebellum and of the spinal cord on the opposite side.

The course of events appears to be this ; first, there is, as the result of chronic inflammation of the meninges, or of the cortical substance, wasting of one side of the brain. To compensate for this the skull becomes thickened, and serum is poured out beneath the arachnoid and into the ventricles. Then, since those parts of the brain which are connected with motion are wasted, the limbs whose action is governed by them are imperfectly nourished and become atrophied.

With reference to the microscopical appearances found in atrophy of the brain, a case is quoted by Dr. Major in the *Journal of Mental Science* for 1879, in which especial attention was paid to this point, with the following result.

The cortex of the convolutions of the affected side was seen to be reduced in thickness, and under a low power seemed to be made up of small round cells of uniform size, here and there only a pyramidal corpuscle being observable, in marked contrast with the appearances in the cortex of the opposite hemisphere. Examination with higher powers showed that the nerve-cell elements were extremely few, as well as small, ill-developed, and deficient in branches. The intercellular matrix of the neuroglia was denser and coarser than in the right hemisphere and under normal conditions. In the internal white matter the nerve fibres were nearly absent in some places, the whole tissue being represented by numerous Deiter's connective tissue cells with their dense intercommunicating network, naked nuclei and occasional vessels. The atrophied lobe of the cerebellum showed a condition of thickness and wasting of the outermost grey layer, few and ill-developed cells of Purkinje, and excess of connective tissue corpuscles.

The spinal cord was not examined by Dr. Major, but was so by Dr. Taylor in a case related by him. There was shrinking of the anterior grey cornu and of the antero-lateral column of the affected side, but the number and size of the ganglion cells of the anterior cornua were almost identical. The cells on both sides were identical in colour after staining, in structure, outline, and processes, with those of the opposite healthy side. There was nowhere any degeneration of the nerve fibres in the white columns, and the dorsal and lumbar regions were distinguishable in no respect from those of a healthy cord.

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