I DESIRE under this title to place certain cases of diffuse sclerosis in which the cortical layers of the cerebrum are the parts primarily or principally affected, and where the microscope shows the presence in these layers of a homogeneous ground substance, or of a finely fibrous network with a few spider-cells in the meshes, occupying the position of the neuroglia. In children but few cases in which this form of disease exists have been accurately described, and in our text-books the whole subject of diffuse sclerosis in children is still obscure.

That the form of cortical sclerosis or degeneration which I am about to describe, can be, in many cases at least, clinically differentiated from other forms of cerebral sclerosis, I feel confident. From ordinary lobar sclerosis and from the different forms of tuberous and multiple sclerosis, this affection can usually be distinguished. It is, on the other hand, closely allied to the conditions found in adults in general paralysis (paralytic dementia) and in chronic alcoholism.

The cases of this affection of which careful descriptions exist are very few.

1 Read in abstract before the American Neurological Association in Philadelphia, June 5th, 1890.
In the St. Bartholomew Hospital Reports for 1879, Moore gives the case of a girl, five years' old. Previously in perfect health. Six weeks before her entrance into the hospital she fell twice. This was followed by twitching of the left side of the face and hemiparesis of the extremities on the left side. Ten days later occurred sudden right hemiplegia and aphasia. On admission to the hospital the pupils were widely dilated, insensible to light, no strabismus. She could not see, but could hear a little. She resented being touched, frowned and screamed; ground her teeth frequently. Did not control sphincters. Had slight clonic movements of all the extremities, stronger on the left than on the right. No loss of sensation. Shortly after entrance had two epileptic attacks, and later opisthotonos. Died about six weeks after entrance. The autopsy showed sclerosis of the cortex to a large extent; the white matter was unaffected. Microscopically, increase in neuroglia, diminution in number and possibly in size of nerve cells.

Schmaus' case is somewhat analagous, but we are inclined to think should rather be classed among the lobar sclerosis, in which the process may be very diffuse. It occurred in a girl three years old. The pathological condition was "Diffuse sclerosis with enormous atrophy of the cerebrum, especially of the convolutions. Hydrocephalus internus et externus ex vacuo. Pachymeningitis interna. Hydrocephalus externus. Atrophy and sclerosis of the anterior columns of the cord, microscopically. All cortical layers diminished, but easily distinguished. Principal change is in the existence of a very thick net-work of fine fibres, lying between the ganglion cells. The ground substance in the white matter is likewise a fine net-work."

My own case, which was quite remarkable in some respects, is as follows:

The patient, when first seen, April, 1887, was a boy 13 years old, a native of Boston. The family history, which was carefully enquired into, showed nothing remarkable. No neurotic tendency.

The patient was born at full term, the labor was natural, and no instruments were used. His general health has
always been excellent. He learned to walk when 19 months old. Has always been bright and intelligent, wrote a good hand, and at the time of his accident was in the 5th class of the Grammar school.

In August, 1886, he had a fall (was said to have been knocked down) and struck his head against the curbstone, making a wound over the R. parietal bone, the scar of which still remains. He did not, however, complain of any pain in it, though it was somewhat tender, and his whole head has been sensitive all winter. At the time of the accident, he was perfectly healthy, and he was promoted in school after the injury, but his mother believes that after it, he was stupid at school, and did no work.

The first thing peculiar noticed about the patient was three weeks after the accident, when he forgot the baby whom he had taken out of doors. He was accustomed to take the child out, and was very fond of him. Two months later, he lost himself one day, and could not be found until the day following, having spent the whole night out. He could not tell people where his home was, nor find it himself.

Since the accident, he has gradually lost his memory, become forgetful and lost interest in external objects.

In the beginning of his illness it was noticed that he kept his tongue in constant motion in the R. side of his mouth, but this ceased some time ago. He has become awkward on his feet and cannot use his hands, as he used to do.

Physical strength and general health, excellent. Has had apparently slight difficulty in sight.

Present condition (April, 1887): Physically well developed and in good condition. Is very forgetful and childish, weak-minded. Will come across the room to see a watch, and smiles at it like a baby. Sits quiet much of the time with a vacant expression. Understands fairly well what is said to him. At home, likes to sit by the window and watch what goes by. Can read the name on a wagon, but does not connect any idea with the words, and cannot even do this now as well as he could at first. Remembers the boys who
were in his room at school since his injury, but nothing of those who were there previous to his injury. Speech slow and indistinct. Occasionally omits words. Never uses the wrong word. Can use neither hands nor feet well, now. 

Physical Examination: Of average size. Muscular development good. Head of normal size and shape. Scalp 1 1-2 inches long over R. parietal bone. R. abducens seems weak. Tongue protruded straight, but seems to move more readily to L. Cannot be induced to squeeze with hands but can pull fairly well. L. hemi-paresis. Clonic spasm of L. arm and face. Incoordination of L. upper extremity and probably of L. lower extremity. Stands with legs apart, as in hypertrophic paralysis. Waddles in walking, which is difficult. Sensation is possibly diminished, as he is said not to complain of burns or wounds on hand. He declares, however, that he feels the pricking of a pin everywhere on the legs readily, and says "prick" when he is touched with it. Skin reflexes on trunk and plantar reflexes excellent. Knee-jerks alike, exaggerated.

On the 24th of April, entered the Carney Hospital. His eyes were examined by Dr. Standish, who reported R. eye veins tortuous, exudation along the arteries and atrophy of choroid. L. eye vessels all small. Neuro-retinitis of both eyes, most severe on the right.

Child seems happy. Has a habit of repeating the emphasized words of a sentence spoken to him, and also repeats his own words. Temperature morning and evening, normal.

At this time he was seen in consultation with Dr. Burrell, and it was decided to trephine at the seat of injury in the hope of finding some source of irritation, it being evident from the child's condition that he was constantly growing worse, and the prognosis being distinctly unfavorable, unless some change could be induced shortly.

On the 27th of April, therefore, the patient was etherized. Slight clonic spasm of L. upper extremity, as patient enters under the influence of ether. An oval, somewhat excavated cicatrix, 1 1-2 inches long by 3-8 inch. in width.
is found externally, situated 3-4 inches to the right of the sagittal suture, and somewhat posterior to the line passing from one auditory meatus to the other over the vertex.

The cicatrix was probably over or just posterior to the median portion of the ascending parietal convolution.

The skull was trephined by Dr. Burrell over the cicatrix and a large disc removed. The dura-mater appeared normal, and there was no bulging. A trocar inserted forwards and somewhat inwards, met with no resistance. The bone was replaced. The external wound was closed with cat-gut sutures. The wound healed readily, and the patient was discharged from the hospital.

There was a slight, temporary cessation of the clonic spasms, but no permanent improvement. The only change noted while in the hospital was, that he lost the power of speech almost entirely. Screams considerably.

On the 22d of June, two months after the operation, I saw the patient at his home. He was found lying on a mattress placed on the floor. He is unable to get up by himself, but when standing can walk from one room to another. After first leaving the hospital, he screamed almost constantly for two days; now screams at intervals, if at all excited, and sometimes without apparent cause. Cannot talk at all now. No change in the special senses noticed. There has been, since April, an increasing incoordination and paresis, involving all the extremities, but more markedly those on the L.

August 8, 1887. On the whole, no worse than at last report. Does not scream much, except when changed after getting clothes. There has now been incontinence of urine for some time.


November 7. L. side now (completely) paralyzed.
Shortly after this, being utterly helpless, and his mother no longer being strong enough to lift him, was committed by Dr. Jelly and myself, and sent to the Boston Lunatic Hospital. There he died a fortnight or so after admission in a condition of extreme dementia.

The autopsy was performed by Dr. Gannett, and it is published as No. 77 in the Forty-ninth Annual Report of the Superintendent of the Boston Lunatic Hospital for the year 1887.

"Male, 14 years. Autopsy twenty-nine hours after death. Body well developed and fairly well nourished; slight lividity of dependent portions; rigor mortis present. There was a horse-shoe shaped scar over the right posterior parietal region; a circular disc of bone had been removed and replaced, and it had become firmly adherent to the surrounding bone; dura not remarkable. The brain did not quite fill the cavity of the skull, and weighed 1,260 grms.; sulci wide; pia showed a slight degree of thickening and opacity; the meshes contained a moderate amount of thin clear fluid; vessels at the base and fissure of Sylvius showed nothing remarkable. Both lateral ventricles were very greatly increased in size. The surface of the ventricles was quite smooth and quite tough to the sense of touch. Ependyma of fourth ventricle markedly granular. Brain substance in general, pale and very tough. Both nuclei caudati were unusually thin; the other basal ganglia not remarkable; pia readily separable from brain substance. Organ of thorax and abdomen showed no variation from the normal."

**DIAGNOSIS.**

- Wound of earlier trephining.
- Oedema of pia.
- Chronic lepto-meningitis.
- Atrophy of Brain.
- Chronic internal hydrocephalus, secondary to atrophy of brain.
- Chronic ependymitis of fourth ventricle.
- No evidence of compression or obstruction of vena Galeni.
Microscopical examination. Pia showed a slight degree of thickening from growth of fibrous tissue.

Brain.—First layer of cortex showed the finely fibrous network with a few spider cells in the mesh, due to atrophy of the nerve fibres and an increase in the neuroglia. There was a slight degree of nerve cell infiltration of the adventitial sheaths of the blood vessels in the cortex. Beyond this, nothing abnormal was observed.

The interest in this case consists in part in its rarity. Very few cases of primary diffuse cerebral sclerosis in children of this age, have been reported. There are numerous examples of cerebral sclerosis, either congenital or occurring in the earlier years of childhood, but they apparently differ somewhat in character from mine. Pathologically, too, I believe them to be distinct. Secondary cerebral sclerosis due to hemorrhage, tumor, chronic hydrocephalus or other cause (embolism or thrombosis, loss of extremities) is common.

Taking simply Moore’s case and my own as the type of this form, the clinical picture presented is as follows: The onset of this disease was in both cases ushered in by a fall from which the commencement of the symptoms is dated by the relatives. In a varying period after this, there appears some intellectual impairment, shown at first simply as obtuseness, general dulness and loss of memory. At about the same time temporary clonic spasms begin to occur, perhaps first affecting the face or tongue, but gradually involving the extremities, those on one side of the body being less affected than those on the other. These convulsive attacks are for a time unaccompanied by loss of consciousness, though later in the disease true epileptic seizures may occur. Nearly simultaneously with the development of the active motor symptoms, there occurs a loss of motor power, sometimes appearing suddenly as a hemiplegia; sometimes very gradually, but usually involving the extremities on one side especially, later those on the other side being likewise affected. In addition to this loss of power in the extremities, we sometimes find a very marked diminution in the power
of co-ordination or control, likewise somewhat hemiplegic in distribution, which in the earlier stages may even mask or conceal the paralysis. The speech is soon affected, either becoming slow, thick and difficult, or aphasia may occur in connection with a right hemiplegia. The loss of power in the extremities increases as a rule progressively and gradually involves the sphincters, causing incontinence of urine and feces. It has, however, never become absolute in any case yet reported, but has reached such a point that the patient was incapable of sitting up alone or of making any more than a slight motion with any extremity. As the disease progresses the spasmodic manifestations incline to increase in frequency and severity, and we may have epileptoid seizures and opisthotonos.

In spite, however, of the progressive character of both the motor and intellectual symptoms, we find sensory symptoms almost absent. There are no anæsthesiae or paræsthesiae apparent. No pain is complained of and no definite loss of sensation has in any case been determined. Unfortunately we know little or nothing in regard to the condition of the muscular sense. Neuro-retinitis or optic neuritis occurs early. The other special senses seem usually unaffected, but as the progressive failure of intellect becomes more and more marked, no absolute determination of their condition can be made. Like the motor, the mental symptoms progress gradually, but steadily.

In the patient under my care, the mental condition was throughout that of a simple, rapidly progressive deterioration of all the mental powers, a pure dementia, uncomplicated by any intervals of excitement or violence. There was no evidence of melancholia; on the contrary, the patient seemed in excellent spirits, happy and contented, until he reached the more advanced stages of the disease. No hallucinations or delusions were at any time found. The mechanical grinding of the teeth, which became nearly constant at times, and the at least partially unconscious screaming, which in the later stages was sometimes continued for hours, were rather evidences of unconscious or semi-con-
L SCLEROSIS OF THE BRAIN IN CHILDREN.

The symptoms of this affection, in short, are those of a gradually progressive mental deterioration, uncomplicated by hallucinatory or delusional conditions and a gradually progressive loss of power, involving all the extremities, the trunk muscles and the sphincters, a loss of speech and loss of sight, accompanied by the symptoms of motor explosion, tonic spasms, and eventually epileptic seizures; while, on the other hand, we find an almost total absence of all sensory disorders. In my case, we find a very marked co-ordination of the extremities. In neither of these cases was there either tremor or nystagmus, both of which were present in the case of Schmaus.

A case related by Turnbull and by him placed under the title of general paralysis, commencing in a boy of eight, presents symptoms very similar to those of diffuse cortical sclerosis. In this case there was less apparent prominence of the motor symptoms, although the patient had an attack of hemiplegia, which seemed to act as the starting-point on which the later dementia was engrafted. The duration of the affection in this case was 10 years. At the autopsy was found chronic pachymeningitis, chronic lepto-meningitis, external hydrocephalus, ependymitis and atrophy and flattening of the cerebral convolutions.

McDowell's case in a lad of 18 is clinically very similar. There is no mention of any meningitis at the autopsy, but there was atrophy and induration of the whole of both cerebral hemispheres, and patches of softening in three places.

We now come to the question of diagnosis. Can this affection be accurately diagnosticated during life and distinguished from other cerebral affections, with somewhat similar symptoms?

The most important practical point is to determine in any case whether we have to deal with a diffuse process, or whether the symptoms presented may possibly be caused by some process more or less localized. In certain cases this is by no means an easy matter to determine.
the explosive symptoms predominate and the paresis is slight or unilateral, and especially where, as in my case, a cicatrix exists over the motor region on the side opposite to that of the hemiparesis, the possibility of a localized starting-point cannot be excluded. It is the possibility of a localized lesion or new growth, accompanied by a chronic or subacute inflammation of the meningeal membranes, which sometimes renders an absolute diagnosis impossible. A cerebral tumor uncomplicated with meningitis would be unlikely to produce this combination of symptoms.

In regard to multiple sclerosis, the differentiation from congenital cases and from those forms occurring in epileptic idiots is simple, and may be determined by the history. A number of cases have been reported, however, by competent observers of multiple sclerosis in children, in which the symptoms more or less clearly resembled those of the same disease in the adult, but so far as I have been able to discover, that of Schüle is the only one in which the diagnosis was substantiated by the autopsy. At the present time, therefore, we can only say that we should expect in uncomplicated cases of multiple sclerosis in children a greater similarity to the symptoms of that disease as shown in grown persons than to those of diffuse cortical sclerosis. A slow development, beginning with nystagmus, tremor and paresis, with a distinct subordination, at least until the later stages of the disease, of the mental symptoms.

From the congenital or early forms of cerebral atrophy and from all forms of partial or lobar atrophy (excluding multiple sclerosis) the history of the case is usually sufficient to exclude. But, in any case, the non-progressive character of the disease in these cases forms a distinction.

In cases of primary hydrocephalus, so-called, the symptoms are usually of a very different character from those just detailed. The presence of rachitis would of itself suggest this affection. As a rule the mental symptoms are developed late, if at all, and the symptoms of cerebral compression are prominent at some period of the disease. It is, perhaps, scarcely needful, however, to mention that hydrocephalus occurs as a secondary condition in diffuse sclerosis.
The conclusions that I would draw provisionally are, that there exists a form of diffuse cerebral sclerosis in children in which the cortical layers of the brain are more specially affected, and which is clinically distinguished from the other forms by its appearance in healthy children, either without known cause, or after traumata, by the steadily progressive character of its symptoms, and by the special prominence of the gradually increasing dementia, which finally reaches an extreme degree without a correspondent loss of motor power, and while the sensation is comparatively unaffected. The pathological condition found is pathologically undistinguishable from that found in adults in general paralysis.