# Hemispherectomy for the Treatment of Epilepsy and Behavior Disturbance

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SUMMARY: *This study is based on 28 personal cases. Twenty seven cases of infantile hemiplegia and one case of Stürge-Weber syndrome, with uncontrollable epilepsy and severe behavior disturbance, were subjected to hemispherectomy. Any evidence of a defect in the other hemisphere was a contraindication to operation. In nine cases the left cerebral hemisphere was removed. Following the operation, epilepsy and behavior disturbances were abolished in nearly all of the cases. In those with left hemispherectomy there was no speech disturbance before or after the operation.*

*It is postulated that at birth all cerebral funtions including consciousness, speech, and movement are bilaterally represented, but to unequal degrees. With training and education these functions become more lateralized. Thus, we can explain the greater plasticity of the brain in childhood.*

RÉSUMÉ: *La présente étude est basée sur 28 cas personnels. Nous avons pratiqué l'hémisphérectomie chez 27 cas d'hémiplégie infantile et un cas du syndrome de Sturge-Weber, tous souffrant dépilepsie non controllée et de troubles sévères du comportement. Toute évidence de trouble dans l'autre hémisphére était considérée comme une contre-indication à l'opération. Chez 9 patients l'hémisphére cérébral gauche fut enlevé. Après l'opération l'épilepsie et les troubles du comportement furent abolis chez presque tous les cas. Dans les cas d'hémisphérectomie gauche il n'y eut aucun trouble de la parole avant ou après l'opèration.*

*Nous postulons qu'à la naissance, toutes les fonctions cérébrales, incluant la conscience, la parole et le mouvement, sont représentées bilatéralement, mais à des degrés divers. Avec l'apprentissage et l'éducation, ces fonctions se latéralisent. Ceci expliquerait la plasticité plus grande du cerveau dans l'enfance.*

From the Dariush Kabir Medical School, Tehran.

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I feel grealy honored and, at the same time, humble to be invited to deliver the 1978 Wilder Penfield Lecture. Dr. Penfield was a giant amongst the neurosurgeons of the world. His great contributions to the advancement of knowledge on the functions of the brain and especially in the treatment of epilepsies are known universally. The Neurological Institute in Montreal is his creation as well as his monument.

He was not only a great scientist and a master surgeon, but also a true philosopher. Thus, he wrote "Throughout the analysis of every case, the clinician should be first and foremost a wise physician and an understanding friend to the patient. He should weigh for him the chances of success of surgery and balance this against the best that conservative treatment can promise. He must see the whole problem in the perspective of the patient's own outlook upon life". (Penfield and Jasper, 1954).

My reasons for choosing hemispherectomy as the subject of this lecture are threefold. First, it deals with the treatment of epilepsy in a few carefully selected cases. Second, it throws some light on the physiology of cerebral function. Thirdly, it was the subject of a paper which I presented at the Tenth Middle East Assembly in 1960 in Beirut.

On the subject of hemispherectomy, Dr. Penfield (1954) has written "Among patients who have large areas of abnormality of one hemisphere, abnormality of behavior may appear, together with advancing mental retardation. The behaviour abnormality is often a more important complaint than the seizures themselves . . . Radical complete excision may correct the abnormality of behaviour, may

stop the seizures, and may allow improvement in the mental state. Furthermore, excision of the motor area may improve the voluntary control of the hemiparetic extremities due to a lessening of spasticity".

Dandy in 1928 was probably the first to demonstrate the feasibility of this operation. Dr. K.G. McKenzie of Toronto was the first neurosurgeon to employ this operation in 1938 for uncontrollable epilepsy in a case of infantile hemiplegia. (Williams & Scott, 1939). Krynaw's paper (1950), reporting the operation in 12 cases of infantile hemiplegia with encouraging results, prompted many surgeons in different parts of the world to perform this operation. I have performed the operation on 28 cases, 27 with infantile hemiplegia and 1 case of Stürge-Weber Syndrome.

The 27 cases of infantile hemiplegia were carefully selected. They all had uncontrollable epilepsy and severe behavior disorders. Hemiplegia had occurred in early life from 5 months to 10 years. Their ages at operation varied from 3 years to 25 years, with 24 cases below the age of 15 years.

Hemiplegia was severe, with atrophy of the affected limbs. The hemiplegic side had no bearing in selection of the cases. In 9 cases the hemiplegia was on the right.

All the patients were able to walk. The arm had a good range of movements at the shoulder joint, less at the elbow, and often no movement at the wrist or fingers.

Sensations of light touch and deep pressure were present on the affected side. Sensation of pin prick was usually well recognized, but often did not cause pain. Two point discrimination was usually faulty. Joint movements were usually perceived in proximal joints, but were absent in the small joints of toes and fingers. Before the operation it was often difficult to examine sensation in these patients because of their behavior disturbance.

All cases had major attacks, some of them had episodes of status epilepticus. Jacksonian attacks were frequent. Most patients were unmanageable at home and none could be educated at school. Destructive tendencies, rudeness to relatives and strangers, and attacks on other siblings were common. There was some evidence that memory was not affected as they often knew all the profane words in and out of dictionaries — and these they used freely on their medical attendants! Some of the patients could recite poetry which they had memorized.

## INVESTIGATIONS

As the operation can benefit only those who have one normal cerebral hemisphere, all patients had both hemispheres examined.

Speech was always examined with great care. With unilateral lesion occurring in infancy, speech may be delayed for a few months if the "dominant" hemisphere is damaged, but it soon becomes normal.

Plain x-rays of the skull showed the affected side of the cranium was smaller than the opposite side; the inner table was smooth and the cavarium was thicker. Pneumoencephalography showed marked dilation of the lateral ventricle on the affected side, with deviation of the ventricular system to that side. Large arachnoidal cysts were often seen on the side of the lesion. (Fig. 1).

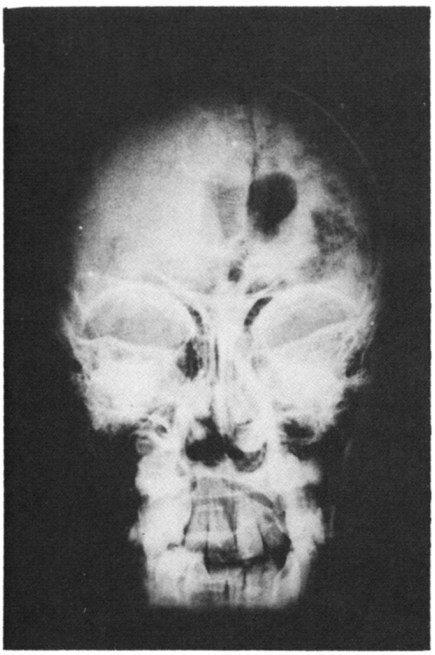

*Figure 1* — Pneumoencephalogram. View demonstrating enlarged left lateral ventricle with deviation of the ventricular system to the same side. Calvarium is thicker on the left side with smooth inner surface.

These features can be seen on CT scan as shown in Figure 2. Even slight enlargement of the ventricle on the

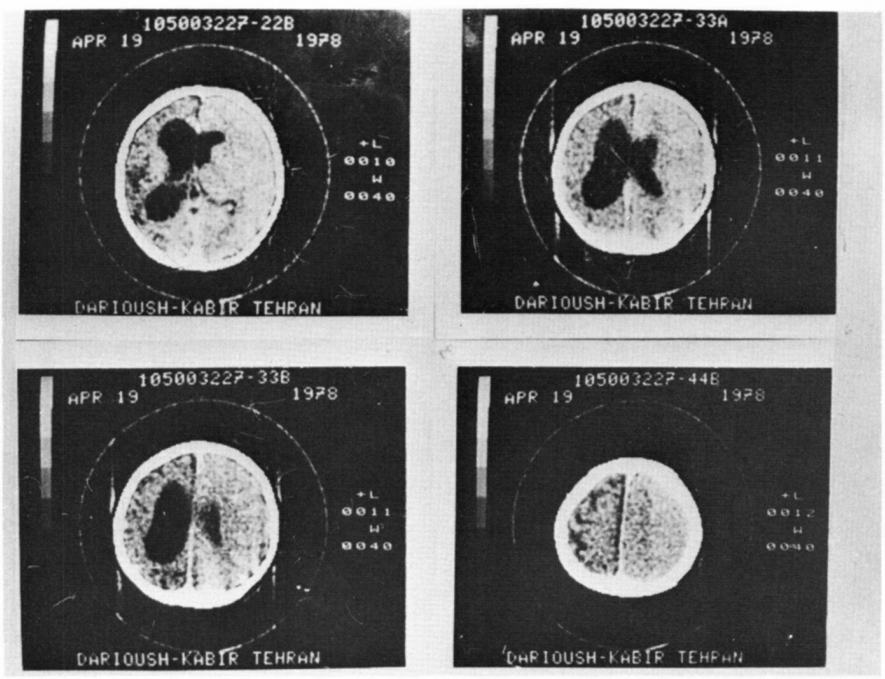


*Figure 2* — CAT in a patient with right infantile hemiplegia. Large left lateral ventricle with large subarachnoid spaces and deviation of the ventricular system to the left side and a much smaller left hemisphere should be noted.



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"normal" side is a contraindication to operation. EEG was usually grossly abnormal with the maximum abnormality on the affected side. Angiography often showed narrowing of the middle cerebral artery (Fig. 3).

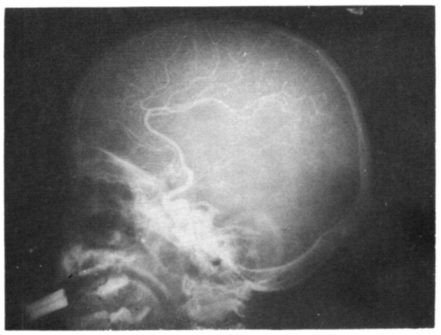

*Figure 3* — Lateral angiogram of a patient with right hemiplegia, showing severe narrowing of the Sylvian arteries.

## OPERATION

In most cases the dura was found to be thicker than normal with subdural adhesions. In two cases there was definite evidence of absorbed infantile subdural effusions. The surface of the brain was usually covered with thick white material. Large subarachnoid pools were present (Fig. 4). Cortical atrophy was marked and the gyri felt hard. In three cases the hemisphere was removed in one piece but others were removed in 3 or 4 pieces. Basal ganglia, except for the head of the caudate nucleus were left intact.

## MORTALITY AND COMPLICATIONS

We had two deaths in the first 8 cases. There has been no operative mortality in the last 20. We had two cases of hydrocephalus, collection of fluid under pressure in the empty half of the cranium, but these made excellent recoveries with shunts. Three cases had pyrexia lasting 1 to 2 months.

## RESULTS OF THE OPERATIONS

As soon as the patient regained consciousness he was able to move his paralysed limbs and could speak. the character and range of movements and speech were the same as before the operation. Once the patient became ambulant, improvement in gait and the range of movement in the affected arm could be seen.

In most cases there were no further seizures and no medication was needed. In one case there were 3 fits and small doses of anitconvulsants were prescribed, with no recurrences. In two cases there were attacks of peculiar sensation in the affected limbs, with no abnormal movement or loss of consciousness.

Behavior gradually improved. Often patients became keen students, as well as polite and docile. In patients with left hemispherectomy arithmetic ability remained poor.

It has been suggested that some of the improvement was due to withdrawal of anti-epileptic drugs, but I think this played a small part.

There was no change in sensation. Sensation in the affected limbs varied from patient to patient. In all patients the sensations of touch, pain, and position were maintained, but, with careful examination, differences on the two sides could be seen. For instance, position senses were better preserved for proximal joints. Streognosis was always absent. Two point discrimination was often abnormal but in some patients was nearly as good as on the normal side. After the operation all patients had contralateral homonymous hemianopsia. The EEG became practically normal with lower amplitude on the operated side.

For geographical and sociological reasons we were unable to trace eleven patients who lived in remote villages. Some of these might have died from late complications. Wilson (1970) analysed 50 cases operated by McKissock and found that 16 patients had died, some of them 10 years or more after the operation. The commonest cause was intracranial hemorrhage.

## NEUROPHYSIOLOGICAL IMPLICATIONS

Studies of these patients before and after the operation pose a number of questions. These can be divided into movement, sensation, behavior, and epilepsy.

## MOBILITY

Paralysed limbs before the operation had a certain amount of power and voluntary movement which was uniform in range and character in all the patients. This same mobility was present after the operation. There-

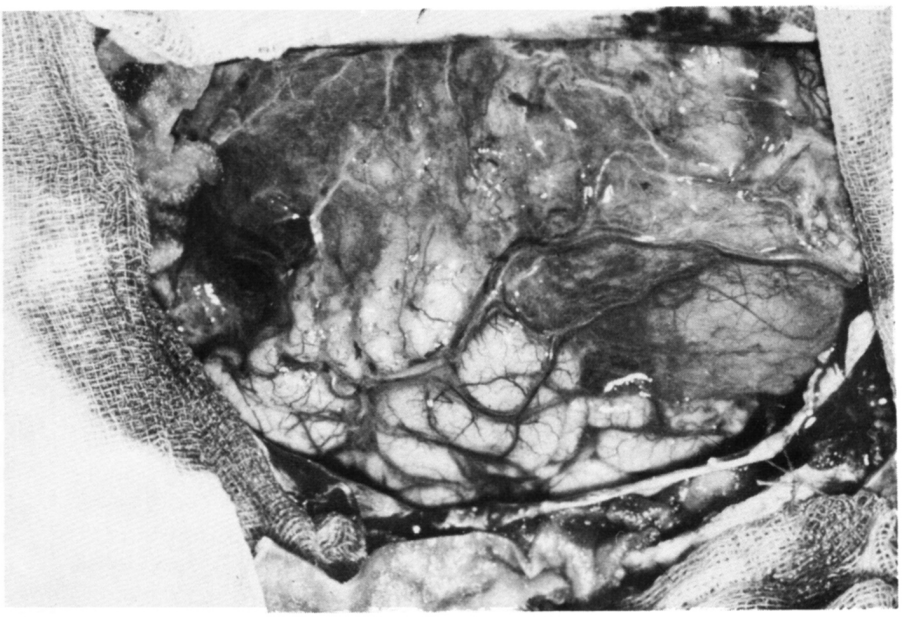

*Figure 4* — Surface of the brain as seen at operation. Large subarachnoid pools and atrophy of the gyri should be noted.



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fore, the diseased hemisphere plays no part in these movements, and the question is: Where are the movements initiated? Bastian's conception of plasticity of the brain (1880 and 1898) and Foerster's theory of synergies (1936) do not really explain this phenomenon, but both of these authorities emphasize that synergies and functional plasticity of the brain are greatest in children.

Penfield (1954) put forward his theory of a centrencephalic system, and believed that function does not originate only in the cerebral cortex. I quote: "Skilled finger movement, like that of a pianist, can only be elicited from the precentral gyrus when it is played upon by well integrated impulses that come to it in a normal manner from some source within the brain that is functionally higher than the convolution itself. When he begins to play, the pianist does so with both hands and it would seem that only from the upper brianstem could come the directing impulses that cause the fingers to move across the keyboard, so skillfully reproducing the melody."

Luria (1966) in discussing Hughling Jackson's three levels of function (1931) and the functional system proposed by Anokhin (1940) stated that the individual areas of the cerebral cortex cannot be regarded as fixed centers but rather they are staging posts or junctions in the dynamic system of excitation in the brain and that these systems have an extremely complex and variable structure. He considered that the recovery of a function must not be attributed to a new vicarious center but rather to a structural reorganisation into a new dynamic system widely dispersed in the cerebral cortex and lower formation. He suggested also that the higher mental functions including speech, resulted from the combined activity of both cerebral hemispheres, each making its own, though not equal, contribution.

Zülch (1978) in a recent paper on hemispherectomy, believed that the mobility of the limbs in hemispherectomised patients was due to the function of mesencephalic structures. In support of this he suggested two arguments. He believed that movements in hemiparetic hands and newborns originated in the mesencephalon, and with progression of myelination higher voluntary function becomes more cortical superceding the mesencephalic movements. The second argument is that in patients with gunshot wounds of the brain when motor areas of both sides adjacent to the sagittal sinus are damaged, and also in patients with Little's disease, they are still able to walk, though imperfectly.

However, in the examples cited by Zülch the damage was inflicted to the areas where no fine movements were initiated. Hughling Jackson (1931) in an analysis of his patients showed that destructive lesions of the areas of the hand and fingers caused much more permanent disability than in the area devoted to the legs. Bastian (1898) had the idea that plasticity of adjacent areas of the brain could replace some of the functions of a damaged area of the cortex. Penfield (1954) has shown that the motor cortex is not a geographical area with rigid boundaries, and movement can be elicited from areas both anterior and posterior to the so-called motor cortex. Brodal (1973) in observing his own handwriting, after a stroke affecting the left side writes ". . . that the motor disturbance in handwriting is most likely due to the interruption of some corticofugal fiber systems which act on the motoneurons of both sides." After styding our patients, it is further suggested (Ameli, 1963) that there is bilateral representation of coarse movements in the brain of animals as well as man. In man and non-human primates who use their hands for fine movements lateralisation is much more developed. In lower mammals the pyramidal tracts are not as well developed as in man. We have observed that hemispherectomised rabbits show very little defect in movement.

For many decades neurologists have known that in hemiplegia most of the movements which are bilaterally simultaneous are preserved. These include movements of the upper part of the face, eyes, tongue, trunk, etc. Highly specialized movements of the fingers and hands are maximally affected.

There is evidence that movement of a limb can be initiated from the ipsilateral himisphere.

Bates (1951) demonstrated that stimulation of the medial surface of the human brain after hemispherectomy produced ipsilateral movements. Luria (1966) noted in gunshot wounds of the right hemisphere there were some disturbance of functions in the right hand.

Researches of Sperry (1968 & 1970) on complete section of the corpus collosum for the treatment of uncontrolled epilepsy demonstrated that a cerebral hemisphere can initiate movements in the ipsilateral hand. In these patients, if a signal is flashed for one tenth of a second on the right visual field, which is represented in the left occipital cortex, the message is received by the left hemisphere alone, and the right hand chooses the object indicated by the message. In some subjects the left hemisphere can, to a lesser degree, bring about purposeful movement of the left hand.

Zülch (1978) showed an interesting post mortem of a porencephalic child in whom the medulla oblongata revealed only one pyramid. This supports the idea that motor impulses to both sides may travel through the one existing normal hemisphere.

It seems that like speech, in early life, bilateral representation is much more evident.

One of our cases subjected to hemispheectomy was a girl aged 8 wth Stürge-Weber syndrome, uncontrolled epilepsy and severe behavior disturbance but no hemiplegia. After operation she developed a complete left hemiplegia, which gradually improved within 6 months. Seen 6 years later she was able to walk and also use her left arm for some functions. Mentally she was normal; she had become literate and was studying. In this patient the ipsilateral brain was able to produce certain movements which were of gradual development.

## SENSATION

There was no change in sensation after the operation but the range and refinement of sensation varied from case to case.

Zülch (1978) reported that the patient considered to have the best sensory performance in the 26 cases


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studied had perception of light touch, deep pressure, topical pattern pain, temperature, graphesthesia, position sense, vibration, and two point discrimination. In one of his cases, two thirds of the thalamus had been removed and the child had good sensory performance.

## SPEECH

Speech is one of the most highly specialized functions of the brain and is unique to the human species. In infantile hemiplegia with speech disorder usually both hemispheres are damaged. We investigated a few of these cases and found that the right lateral ventricle was slightly to moderately enlarged and there was marked enlargement of the ventricle on the left side. Wilson (1970) stated that 6% of his cases had permanent post operative speech disturbance, and all these had gross mental defect before the operation.

Therefore, when speech is affected or there is evidence of mental defect the patient is not suitable for operation.

In nine of our cases with left hemispherectomy, immediately after the operation the speech was normal, demonstrating that the right hemisphere was performing this function before the operation.

Penfield (1954) observed that vocalisation and arrest of speech can be elicited on electrical stimulation of either hemisphere.

Smith and Sugar (1975) in studying a case 21 years after left hemispherectomy found that the patient's language and intelligence had improved and they stated "This case indicated that in left hemispherectomy for early brain insult, the right cerebral hemisphere and other intact structures can furnish the necessary neuroanatomic substrata for the development of superior language and verbal reasoning skills."

Luria (1966) on the same subject writes that ". . . the degree of dominance of one hemisphere in relation to lateralised processes such as speech varies considerably from case to case and this factor introduced a considerable element of diversity into the local pathology of higher cortical functions".

We think that this diversity depends a great deal on training and refinement of function.

In comparing 50 cases of illiterate patients with 50 literates with comparable lesions we found that lesions of the left hemisphere in the first group caused much less disturbance of speech and they recovered their speech sooner and to a greater degree (Friooz-Bakhsh, 1977).

## EPILEPSY

In a majority of cases, if carefully selected, seizures were completely abolished. There has been no evidence of development of secondary foci in the other hemisphere. Two of our cases had minor sensory attacks in the paralysed limb postoperatively. These probably originated in the thalamus.

Walker (1955) noted focal attacks for several days after the operation in a hemispherectomised patient. This prompted him to inject penicillin in oil into the basal ganglion of monkeys resulting in focal or psychomotor attacks in most of the animals. This does not negate the epileptogenicity of the ipsilateral cortex, but it emphasizes the potential of the basal ganglia.

## BEHAVIOR DISTURBANCE

All cases showed marked improvement in behavior in a few days to a few months after operation. The parents' attitude and their co-operation as well as the social environment played an important part in the outcome.

Wilson (1970) found that behavior disturbance was abolished in 50% and improved in 40% of cases.

The improvement may be due to the absence of abnormal stimuli from the diseased hemisphere. These stimuli probably interfere with the integration of all the impulses received by the organism. Without this integration, normal thinking, formation of ideas, and social behavior are not possible.

In conclusion, I suggest that at birth all cerebral functions are bilaterally represented, but not to equal degrees. With training and education lateralisation of fine movements and sensation as well as speech become more obvious.

The higher this refinement develops and the longer it is continued the more localised the function and the more severe and permanent are the results of injury. This is probably the reason for the greater plasticity of the cerebral cortex in childhood.

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