

**SUBTOTAL HEMISPHERECTOMY IN TREATMENT OF EPILEPSY.**

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There are many children suffering from cerebral palsy and epilepsy who need institutional care, and they are a burden to the society and a source of unhappiness and anxiety to their parents. A small number of this group if selected carefully will benefit from the operation of subtotal hemispherectomy.

Dandy (1928) was probably the first to demonstrate the possibility of subtotal hemispherectomy but it was Mackenzie who first performed the operation for this type of case in 1938.

In 1950 Krynaw published his encouraging results in 12 cases of infantile hemiplegia with epilepsy and behaviour disorders. Since then a number of cases probably more than 400 have been submitted to this operation in different parts of the world, and on the whole Krynaw's original optimistic views have been upheld. Nevertheless the value of subtotal hemispherectomy has not been sufficiently recognised.

To-day I will discuss 22 cases who have had this operation in our service in the last 7 years.

**Selection of patients.**

As in many major surgical procedures to obtain the best results cases have to be carefully selected. The most suitable patients are those who have the triad of,

1. Hemiplegia.
2. Uncontrollable fits.
3. Behaviour disturbance.

Age is no barrier to selection. The youngest in our patients was 3 and the oldest 25 years: Hemiplegia must have occurred in infancy.

It is necessary to prove as far as possible that the damage is confined to one hemisphere. Hemiplegia in all our cases except one (vid. infr.) was very marked. The upper extremity is usually much more

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affected, but both upper and lower limbs are smaller than the normal side.

There is no useful movement in the affected hand, which is deformed and held in extreme flexion at the wrist. Fig. 1 & 2. The side affected is not important. We have removed the left hemisphere in five cases, with no effect on the patient's speech. Epileptic fits varied in number from one a month to many a day and six of the patients had had one or more episodes of status epilepticus.

Behaviour disorder varied from case to case, but all of them were incapable of being educated. Only two of these patients had reached the second class and no further. Most of the patients were unmanageable at home and on many occasions parents wished to have the child "either killed or cured."

Destructive tendencies and rudeness to relatives and strangers were the most frequent complaints.

One curious phenomenon which was present at least in half of our cases was an enormous vocabulary of profane words, which they used on their medical attendants.

#### Pathology.

In twelve cases there was history of fever immediately preceding the onset of hemiplegia. In one case it was typhoid and another meningitis, but in others there was no indication to the nature of infection. In one case the middle cerebral artery was very small and in another there was calcification in the wall of the artery. It is possible that the cause of brain damage in a number of cases is arteritis of the middle cerebral artery. This needs further study of cerebral vessels both as regards angiography and histology. In two cases the operation findings suggested absorbed subdural hematoma.

#### Investigations.

For selection of cases as already stated it was important to demonstrate that the brain on the other side is normal. All the cases were submitted to plain x-rays of skull, pneumoencephalography, and E.E.G. Some cases also had angiography. Straight x-rays often shows the affected side is smaller than the normal with no convolitional markings, with thickened calvarium, elevation of petrous pyramid. (Dyke — Davidoff. Masson Syndrome). Fig. 3 & 4

Pneumoencephalograms show the lateral ventricle on the affected side enlarged. and displaced to the same side, with displacement of the 3rd. ventricle. Fig. 5 & 6.

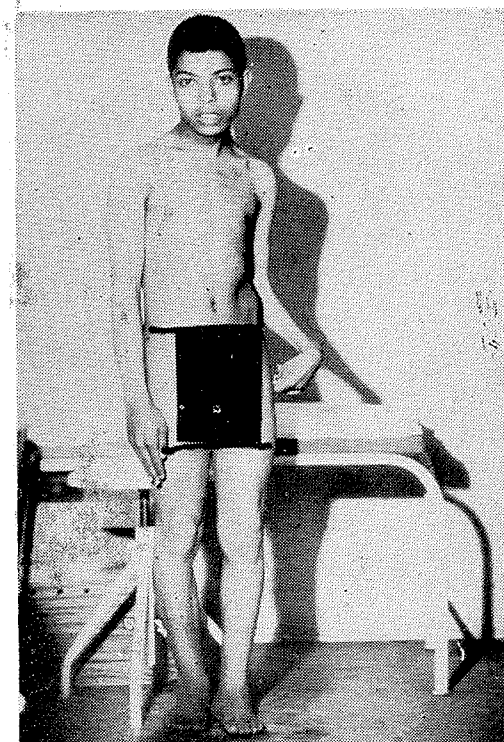


Fig 1

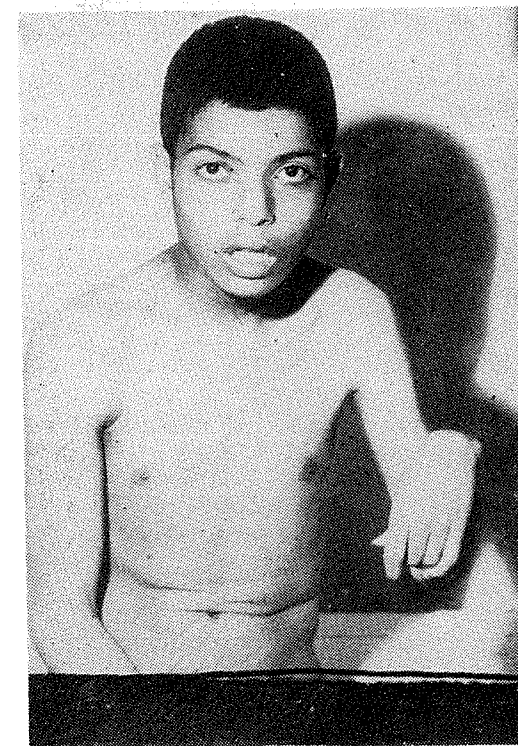


Fig 2

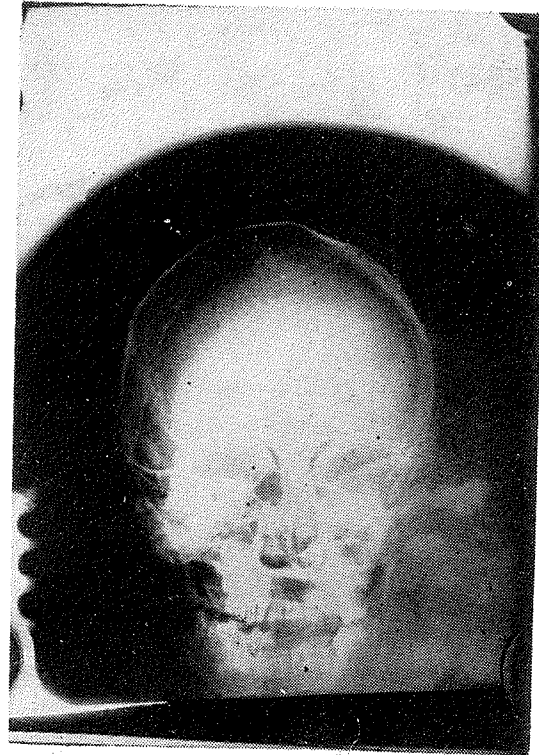


Fig 3

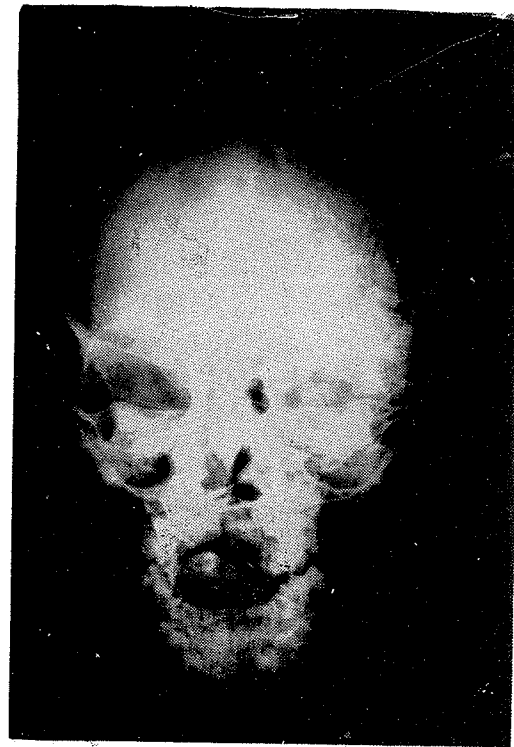


Fig 4

This does not mean that there is hypertrophy of the normal hemisphere. What actually happens is that the normal growth of the brain which usually causes enlargement of the head, in these patients can use the available space on the damaged side. That is to say that as there is room in the affected side some of the growth of the normal hemisphere is accommodated by the available space.

Large subarachnoid pools and porencephalies are often seen. Fig. 7

If both ventricles are enlarged then the case is not considered suitable for operation.

E.E.G. shows predominance of abnormality on the affected side. Fig. 8).

In a case of sturge Webber syndrome with uncontrollable epilepsy and severe behaviour disorder with very slight hemiparesis this operation was performed. Fig. 9 & 10.

Mental behaviour very much improved. There was no epilepsy post-operatively, but she had severe hemiplegia.

#### Operation.

Intratracheal general anaesthesia was used in all cases. In 15 cases moderate hypothermia was also employed. A large osteoplastic flap is turned down. In most cases dura was abnormally thick. In one case it measured 3mm. In one case there was subdural ossification, which we thought to be an aftermath of an absorbed subdural hematoma. Another case had a large subdural hygroma with thick capsule. Brain surface is often covered with thick white arachnoid, not unlike a film of ice cream. This is mostly seen over the central part of the hemisphere. Large subarachnoid pools are often seen. (Fig. 11)

Markd atrophy of the cortex is always seen, but this is not uniform. Again the area fed by middle cerebral artery is mostly affected. In one case the area supplied by anterior cerebral artery was replaced by scar tissue. Fig. 12.

Brain gyri feel hard to touch. In most cases we have noticed that inferior surface of the temporal lobe is very thin.

Could this play a part in behaviour disorders?

To remove the hemisphere we start by dealing with the middle cerebral artery. This is easily found by opening the thick arachnoidal adhesions in the fissure of sylvius, and following them to the carotid artery, a silver clip is applied to the artery near its origin. In one case referred above the artery was calcified and we had to crush it first with the aid of a pair of artery forceps before applying the clip.

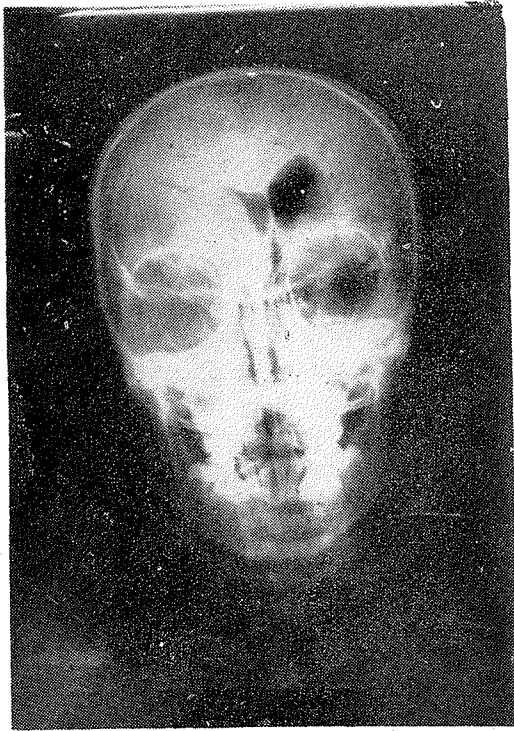


Fig 5

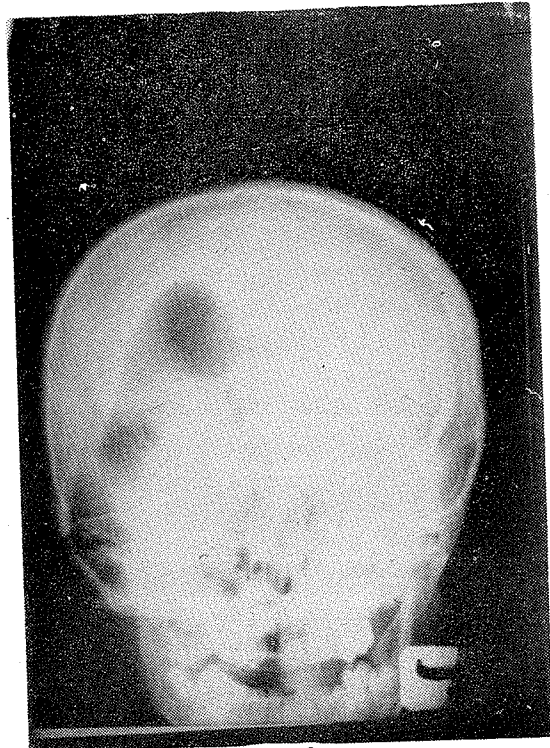


Fig 6

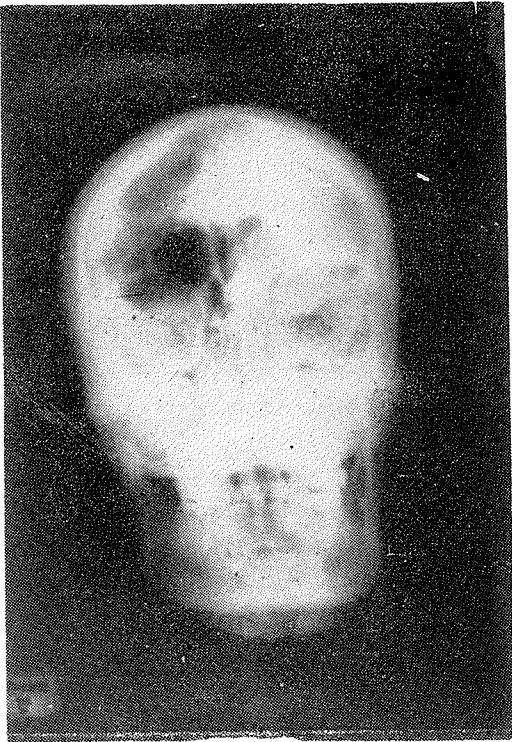


Fig 7

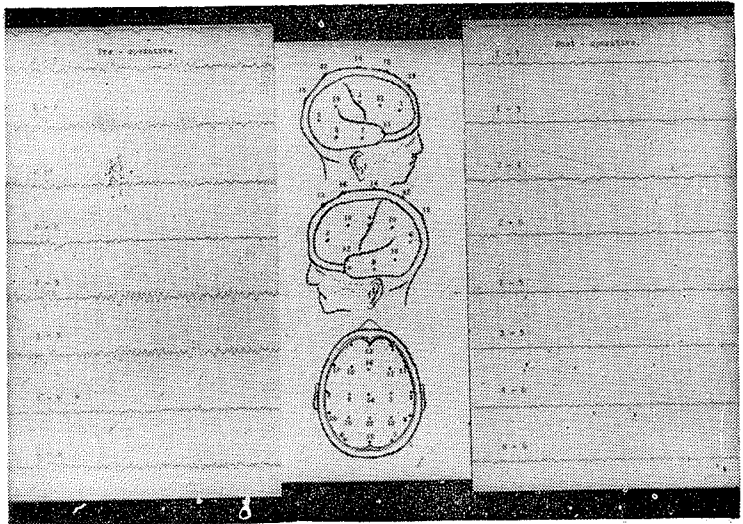


Fig 8



Although the hemisphere can be removed in one piece and we have done this on two occasions (Fig. 13) we prefer to remove the hemisphere in 3 or 4 pieces.

A vertical incision is made into the ventricle, and a cottonoid is applied to the foramen of monroe and then the lobes are removed from the inside of the ventricle, as well as from the outside. Temporal lobe is removed first and then the frontal and finally parietal and occipital lobes together.

Foramen of Monroe has to be carefully plugged throughout the operation so that no blood enters the 3rd. ventricle. Veins draining into the sagittal and lateral sinuses have to be carefully clipped or coagulated near the cortex. Further bleeding can be easily controlled with gel foam. We remove the cortex of insula and often a part of caudate nucleus. Otherwise the basal ganglia are left intact.

The choroid plexuses are not coagulated, and we believe production of C.S.F. from undamaged plexus would fill the empty space and keep the balance of pressure on the two sides.

One of the post-operative dangers of this operation, mid brain shift to the operated side, in this way is minimised.

We have had. no "unilateral hydrocephalus" as is alleged to occur if the choroid plexus is left intact.

We plug the foramen of Monroe with a piece of gel foam in the hope that blood stained fluid does not enter the 3rd. and 4th. ventricle. The cavity is filled with saline and the wound is closed in layers. We have found that closure of the wound without drainage gives a smoother post-operative course.

#### Post-operative Complications.

Unfortunately we had 3 deaths in this series. Two of these were avoidable. One case died from meningococcal meningitis. At the time we had an epidemic in Tehran. The other patient died from a staphylococcal meningitis, two months after operation. For a time we thought we could save the child, but empyema was formed in the cavity and even removal of the bone flap did not help. The 3rd. case died 36 hours after the operation without regaining consciousness.

Three cases had long drawn out slight temperature, with drowsiness, neck rigidity and Kernig's sign. We could find no organism. When one case was steadily going downhill, we began forced feeding through a nasal stomach tube. He rapidly improved. These cases become so lethargic that feeding may be a great problem. In two cases we had

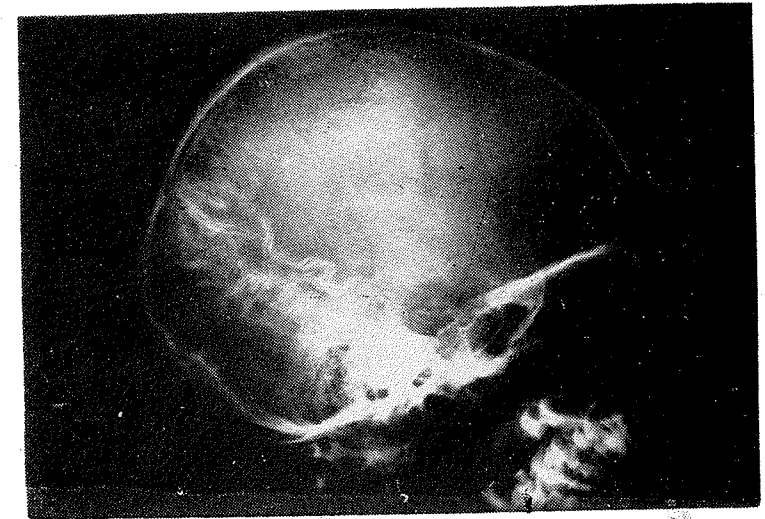


Fig 9

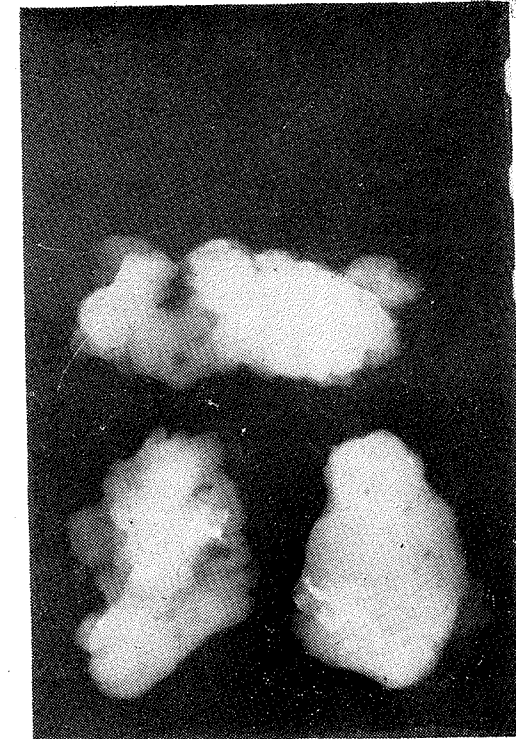


Fig 10

C.S.F. fistula for 2—3 weeks, both these had a scalp drain removed after 24 hours. Since then we close the wound without drainage.

#### Results of Operation.

Our cases have not had long follow-ups, and this is also true of other published series. Our longest follow up has been 7 years. Nevertheless we can give some indications of our results so far. All our cases were mentally very much improved. Case one is now a useful member of the family, and looks after the other children. Preoperatively she could not

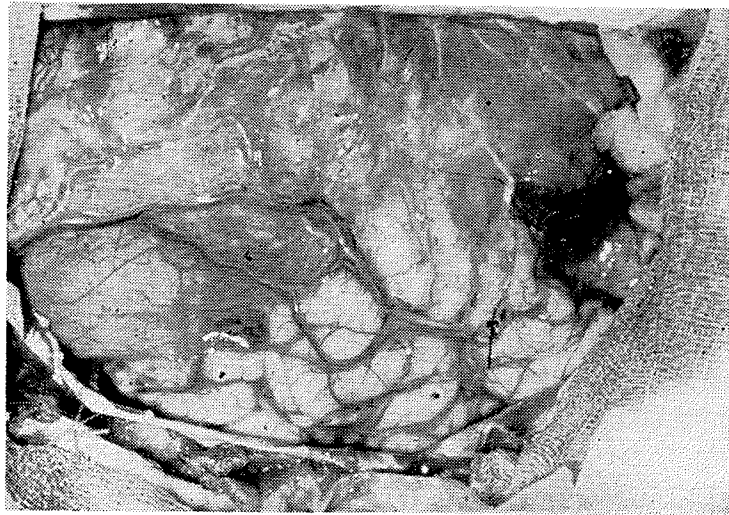


Fig 11

be managed at home or even in the ward, as she was a great nuisance to the other patients.

Case two has resumed his studies that had not gone beyond second class. Case five after one year was in the fourth class. At a medical meeting when he was shown, he complained that he was poor in arithmetic. Case nine who was the oldest of this group (25 years at the time of his operation) is studying hard and wishes to become a teacher.

The number of mentally retarded children from these patients' areas of the country, who are brought for operation and praising the results in the operated cases, are the best indication and encouragement.

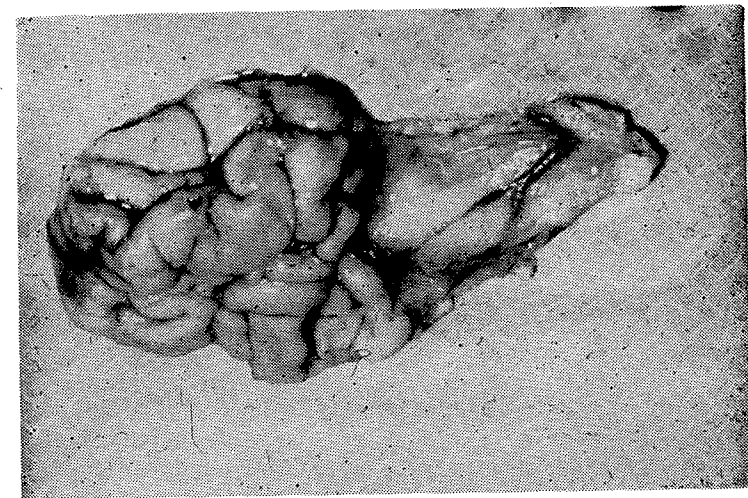


Fig 12

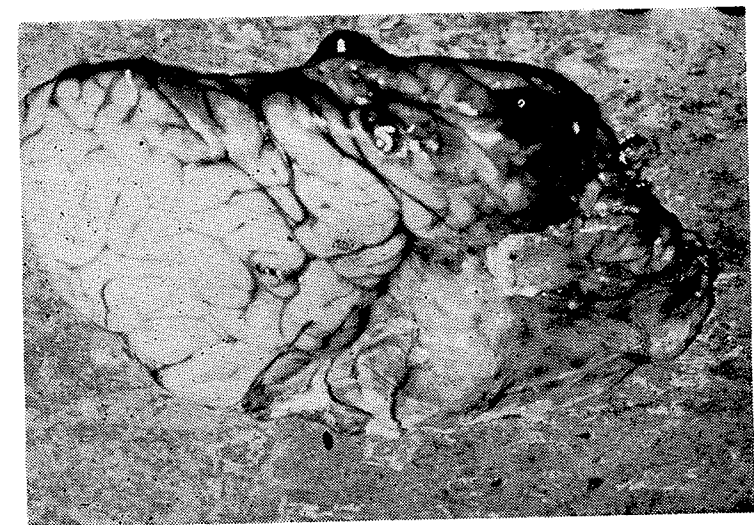


Fig. 13

Hemiplegia is not made worse by operation, and walking is usually improved because of reduction of spasticity. The patients as soon as they regain consciousness can move the affected limbs. This can only be explained by bilateral representation of function in the normal hemisphere.

In all these patients the arm is much more affected than the leg, and the hand is practically devoid of any useful movement and very little developed.

It has been shown that removal of motor cortex in man and animals affects the fine movements of the hand and the coarse movements are unaffected.

Stimulation of medial surface of the brain, hippocampus and fornix (mesopallium) gives bilateral or ipsilateral responses. Furthermore in the lateral pyramidal tracts there are fibres which continue uncrossed to the ipsilateral lower limb.

If we consider all these points together we can understand why

1. With survival of one cerebral hemisphere there is still normal movements in those muscles which normally on the two sides contract simultaneously, as in eyes, neck, chest and abdomen.
2. With destruction of one hemisphere extremities are affected, and the upper extremity more than the lower.
3. Fine movements are lost.

As far as speech is concerned, it is interesting to note that dominance of hemisphere is not definite, but if the dominant hemisphere damaged in the early childhood the other hemisphere can take over the function of speech.

#### Summary.

22 cases submitted to sub-total hemispherectomy are discussed. In selected group of infantile hemiplegia where epilepsy and behavior disturbances are present, and investigations prove that the other hemisphere is normal, this operation is of great benefit. Nevertheless long follow-up of these cases is necessary before a final judgement can be passed. Age should be no barrier to selection, as those of 18 — 25 have benefited as much as young children.

Post-operatively there has been no diminution of movements of the affected side. This has been explained on the basis of recent experiments and observation on the motor function of cerebral cortex.

#### Résumé

22 cas de l'hémisphérectomie subtotale sont discutées

Cette intervention, dans les hémiplegies infantiles où l'épilepsie et troubles du comportement sont présentés, se montre efficace, à condition que l'autre hémisphère soit normale.

Avant de pouvoir donner un jugement définitif un recul de temps important est nécessaire. L'âge n'est pas une barrière au choix de malade, les sujets de 18-25 ans peuvent en bénéficier autant que les jeunes enfants.

Dans les suites opératoires aucune régression de mouvements du côté atteint n'est constaté ceci a été expliqué sur les expériences et observations récentes sur la fonction de cortex cérébral.

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