# CEREBRAL PALSY: THE PLACE OF NEUROSURGERY*

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The object of this paper is to examine the potential role of neurosurgical procedures in the prevention, arrest or rehabilitation of and from a group of maladies included under the somewhat vague and unsatisfactory generic term 'cerebral palsy'. In common usage the term embraces the motor and associated disabilities in individuals suffering from the effects of damage to various parts of the brain.

For obvious reasons most time, money and energy have been directed to the management of these disabilities in children. So much has this been the case that the mere use of the term 'cerebral palsy' has almost invariably been confined to discussions on effects arising from disorders of infancy and childhood. In actual fact 'the prolongation of the span of life, with its increase in geriatric diseases, particularly cardiovascular disease, is rapidly causing a much greater incidence of cerebral palsy in adults than in children'.[1]

## *Etiological Factors and Morbid Entities*

There is some immediate practical value in the classification of cases according to etiology. Tables I-IV, compiled from various sources,[1-4] set out the causes of cerebral palsy in infancy and childhood and summarize certain effects and morbid entities. In all the tables those initial lesions or secondary morbid entities which, to greater or lesser extent, lend themselves to surgical removal are marked with an asterisk.

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TABLE I. BROAD ETIOLOGICAL CLASSIFICATION

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|---|---|---|
| From: | Prenatal Causes .. .. | 20% |
| | Natal Causes .. .. | 70% |
| | Postnatal Causes .. .. | 10% |

TABLE II. PRENATAL CAUSES

A. HEREDITARY OR GENETIC, STATIC OR PROGRESSIVE

Rudimentary pyramidal tracts—Spastic paraplegia

Defects in basal ganglia—Atonic diplegias, tremors, athetoses, rigidities

Defects in cerebellum and/or its tracts—Ataxia

Metabolic anomalies: phenylpyruvic amentia

Ectodermal or mesodermal defects: tuberous sclerosis,* neurofibromatosis,* Sturge-Weber syndrome * and other vascular abnormalities*

Familial cerebral degenerative diseases

B. CONGENITAL NON-HEREDITARY

Erythroblastosis foetalis
Damage to basal nuclei (Kernicterus)
Athetosis and rigidity
Interference with placental circulation
Toxaemia of pregnancy
Maternal asphyxia
Trauma to uterus or foetus
Foetal cerebral haemorrhage, primary or secondary to cerebral softening or/and
Cortical atrophy of various grades or/and
Subdural haematoma(s)

Effects:
Brain cysts and porencephaly *
Regional, lobular or hemispheral atrophy *
Regional, lobular or hemispheral sclerosis *
Microgyria *
Haematoma *
etc.

Maternal infection: neurotropic viruses, syphilis, toxoplasma
Maternal diabetes, liver damage, hyperthyroidism (treatment with thyroid inhibitors)
? Maternal nutritional and vitamin deficiencies
? Maternal gonadal irradiation

TABLE III. NATAL CAUSES

Anoxia: Brain cysts and porencephaly *
Regional, lobular or hemispheral atrophy *
Regional, lobular or hemispheral sclerosis *
Microgyria *
Haematoma *
Meningocerebral cicatrix *
etc.
Mechanical birth injury
Increased fragility of cerebral blood vessels
Bleeding tendency

TABLE IV. POSTNATAL CAUSES

Trauma
Infection: meningitis, encephalitis (e.g. Strümpell-Marie type), brain abscess *
Neoplasm*, often slowly growing
Drugs: lead,* causing anoxia (anaesthetics, barbiturates).
Anoxia from other causes
Arterial occlusions, including acute infantile hemiplegia of obscure origin *
Cerebral haemorrhage and softening following acute infectious diseases *
Venous occlusions

## LESIONS AMENABLE TO DIRECT SURGICAL PROCEDURES

The lesions produced by hereditary or genetic effects seldom lend themselves to direct surgical procedures. These are usually only applicable in instances where the anlage gives rise to some recognizable morbid entity at some time remote from birth. Occasionally tuberous sclerosis occurs in an isolated cerebral lesion which can be removed—the writer has had experience of one such case in an adult woman. An isolated neurofibroma can be removed, but more usually intracranial neurofibromas are multiple and often they are further associated with glioma of an optic nerve or meningiomas.[5, 6] These multiple lesions set a limit to the value of surgery. Isolated and relatively small vascular abnormalities lend themselves to removal or coagulation.[7–10] Extensive removal of abnormal vessels and brain is feasible in some cases of the Sturge-Weber syndrome.[11]

Numerous morbid entities arising from congenital, non-hereditary, natal and postnatal causes lend themselves to surgical removal. Most obvious and satisfactory in the prevention of further development of neurological deficit are the removal of haematomas, the proper handling of head injuries, the treatment of brain abscess. In general, the diagnosis of supratentorial brain tumours in extreme youth does not receive the attention it deserves. Such tumours are not infrequently slow-growing.[1] (*Here an illustration was shown of a fibrillary astrocytoma, which was causing frontal adversive seizures and hemiparesis in a* 3*-year-old child, and was successfully removed.*)

Some of the lesions so far mentioned as amenable to removal, sometimes only in exceptional cases, in themselves constitute a menace to life or can be taken away before they do other irreparable damage. The same does not usually apply to the end-results of a number of causative factors which promote atrophic and sclerotic lesions. Although ablation of these may be feasible it cannot for this reason alone be considered desirable.

### *The Atrophic and Sclerotic Lesions*

This group includes the great majority of the cerebral palsies of infancy or childhood referred to the neurosurgeon. On the basis of radiological studies after air replacement of cerebrospinal fluid, the following gross entities can be recognized:[12]

1. A general dilatation of the ventricular system, sometimes with enlargement of the pericerebral or pericerebellar subarachnoid space.

2. An exaggerated dilatation of part or the whole of one lateral ventricle.

3. Gross cerebral defects, resulting in lobar sclerosis, 'brain cysts' or porencephaly.

Histological studies indicated that the basic alteration common to local microgyria and these grosser signs of brain damage is a characteristic cellular necrosis of the altered cortex.

At the other extreme of this group of atrophies is the local microgyrus.

In clinical practice these morbid entities account for the greater number of *infantile hemiplegias* and *cerebral diplegias*. The bilateral cerebral involvement in the latter at present place them beyond the reach of direct surgical procedures and the attention of neurosurgeons has been concentrated on the first group.

### *Infantile Hemiplegias*

The clinical picture observed in the infantile hemiplegias is slightly variable, but most commonly the child is brought for consultation with a combination of physical and mental defects. There is a spastic hemiparesis, with fixed contractures at wrist and elbow. The affected limbs are usually shorter and generally smaller than their fellows. The skull usually shows an asymmetry. Athetoid movements may occur on the affected side, on which motor weakness and loss of sensory discrimination will be noted. A homonymous field-defect is commonly observed. Mental development varies from bright to very low grade, and behaviour disorders may be most distressing to the parents. To crown all these infirmities, epileptic seizures may occur with great frequency.

The physical disabilities are permanent. The development to adulthood varies, but often there is persistent mental retardation. Epilepsy remains a major disability.

Electro-encephalography indicates a gross dysrhythmia. Ventriculography shows a dilatation of the ventricular system, most often affecting one lateral ventricle in part or *in toto*. There are some cases, however, which do not show the ventricular dilatation and here there is a gliosis of a hemisphere in place of the more usual atrophic, thinned-out, shell covering a lateral ventricle.

(*Two examples were illustrated to show the great variations in the extent of surgical ablations applied to these cases, depending on the extent of the underlying pathology. In the first instance a teenage girl, who had suffered from focal epileptic fits for many years, had a local microgyrus removed with satisfactory result. In the second, a boy of* 19 *years was subjected to hemispherectomy for very frequent major general epileptic attacks, up to* 15 *per day even while on heavy anti-epileptic medication, also with satisfactory result.*)

Various forms of cerebral cortical ablation for epilepsy have been reported in significant numbers, since 1935.[13] The most significant reports have come from Penfield and his associates,[4, 14, 15] who have stressed the epileptogenic role of a zone of apparently normal cortex adjacent to a grossly damaged area.[4, 14, 16]

During the forties Krynauw, in Johannesburg, removed progressively greater areas in atrophic and sclerosed hemispheres in selected cases of 'infantile hemiplegia'.[13, 17] He developed an operation of almost complete hemispherectomy with sparing of the caudate nucleus, internal capsule and thalamus. After a visit to Krynauw's clinic Sir Hugh Cairns introduced the operation to Britain.[18] In 1953 McKissock of London reported on 18 cases of infantile hemiplegia treated by the Krynauw operation.[19] Others in the USA, Spain and Belgium have reported more limited experience with the procedure.[11, 20–23]

In Krynauw's 12 reported cases there was one death and a second case developed an abscess in the basal stump. The others all showed initial improvement in respect of fits and behaviour disturbances,[17] which were considered as epileptic equivalents.[13] In none was the motor disability aggravated by the operation.[17] Perhaps the successors in Krynauw's unit will give us more up-to-date information on follow-up of his cases and tell of their more recent experiences.

In McKissock's 18 reported cases there was one death from infection. The operation eliminated or greatly reduced epilepsy in 16, and removed temper tantrums in 14. Decreased spasticity of affected limbs resulted in greater freedom and facility of movement, but in 2 hemiplegia was made worse—in these local excision would probably have been better advised. Capacity to learn was favourably influenced.[19] In Obrador's 6 reported cases there were 3 deaths, but hemispherectomies were performed after initial failure of more limited resection to alleviate fits.[22]

There were no deaths among the 8 cases treated by hemispherectomy and reported by French *et al.* The results were comparable to those reported by McKissock, but no cases were made worse in respect of motor disturbances.[11]

*The Evaluation of Hemispherectomy*

It is not yet possible to evaluate the full results of hemispherectomy for atrophic and sclerotic lesions arising in infancy and very early childhood. The encouraging signs are the testimony of reported cases with very satisfactory reduction in the number of epileptic seizures up to 5 years after operation, the knowledge that in properly selected cases there follows no added speech or motor disturbance, and that apart from a rather protracted convalescence the operation does no harm, if complications do not ensue. *The indication for the operation is strictly limited to occurrence of frequent epileptic seizures or equivalents uncontrolled by medication in reasonable doses.*

The release of spasticity may facilitate movement,[19] but it may also by virtue of this make orthopaedic deformities more apparent.[11] For the latter reason neurosurgeons are less inclined to perform the operation in the absence of gross infantile hemiplegia, but here again the yardstick must be the frequency of severe ictal episodes.

The effect on intellect and personality may be favourable, but only if the patient is psychologically adjusted to suitable surroundings. Unfavourable social and family relationships, extreme poverty for which there is no remedy, and initial very low intelligence, should be accepted as contra-indications to the operation. Only latent and already existing mental abilities can blossom after the brake of an irreparably damaged hemisphere has been removed. A near-vegetable mentality is made worse; in its presence the convalescence with its necessary re-education becomes a useless torture to patient and attendants and the motor status becomes worse than ever. Such has been my own experience after being persuaded to operate as a desperate measure.

*Regional Cortical Ablations*

The older established regional ablations of meningocerebral cicatrices and limited atrophic and sclerotic areas are still indicated more frequently than hemispherectomy. Again, they are indicated only by the incidence of *frequent ictal episodes uncontrolled by reasonable doses of anti-epileptic remedies.*

The failure of these operations is often due to neglect to remove surrounding epileptogenic, normal-appearing brain-tissue. For this reason very large gross lesions are probably better handled by hemispherectomy, but there is already evidence that nicety of judgment is necessary in making this decision, as witnessed by two of McKissock's cases.[19]

In general it is well to apply to cortical ablations in children the rule that they be done only when seizures are focal and coincide with appropriate clinical neurological motor disability.[24]

*Cervical Arteriovenous Anastomosis*

Attempts at improving the blood supply to brain cells enmeshed in gliotic tissue have been made from time to time, all with singular lack of success. The latest of these methods has been carotid—internal jugular anastomosis in the neck. A large series of cases was reported in 1950, but the results in children with cerebral palsy were disappointing.[25]

*Treatment of Dyskinesias*

Sir Victor Horsley opened up a new field of surgery when he performed motor cortical ablation for athetosis in 1909.[25] The dyskinesias sometimes afford the outstanding disability in cases of cerebral palsy. Hemispheral damage and paralysis dominate the clinical picture in cases due to direct mechanical trauma from various causes, choreo-athetosis does so in cases due to anoxia.[1]

'The surgical procedures performed for the relief of involuntary movements have been varied and at times bizarre, largely due to the fact that the physiological basis of the phenomena were not understood[27]—they are still only imperfectly understood. A variety of procedures have included: intracortical alcohol injections,[28] cortical ablations of parts of areas 4 and 6[29, 30] of so-called area 45,[31] interruption of extrapyramidal tracts by section in the anterior columns of the spinal cord,[32] surgical damage to the dentate nucleus,[33] section of spinal nerve roots,[34, 35] operations on the caudate nucleus and internal capsule.[36, 37] New modifications of some of these procedures are still being elaborated but none can yet be claimed as beyond the 'experimental' stage. Suffice it to say that surgical relief can be given to some cases of *severe* dyskinesia—that 'neurosurgery has been of occasional benefit in certain cases of severe athetosis or tremors'.[1]

## CONCLUSIONS

Various neurosurgical procedures are indicated in a limited number of cases of cerebral palsy of infancy and childhood. It is essential that certain lesions should be diagnosed early and removed, but in the main the role of neurosurgery is confined to the treatment of associated severe epilepsy and less often that of disabling dyskinesias. To obtain salutary effect and not aggravate disability, cases for operation must be carefully selected. For severe epilepsy a range of cortical excisions, from local removal of a microgyrus to hemispherectomy, are available. For dyskinesia operations on cortex, basal ganglia and spinal cord are sometimes suitable.

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