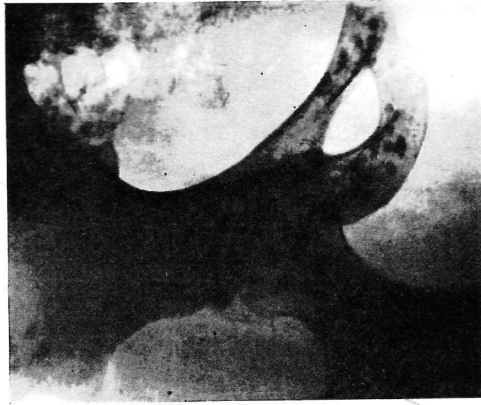


## CONGENITAL DEFECTS

### Morquio's Disease

The vertebral bodies are irregular in size and shape, biconvex, narrower from above down, and with a "tongue-like" projection from the anterior surface (Fig. 381). All the long bones including those of the hands and feet may be affected, and the epiphyses may be irregular in shape and stippled. There are many individual variations.



Left: FIG. 381. Morquio's disease.

Above: FIG. 382. Osteopoikilosis.

### Osteopoikilosis

This condition is sometimes but not always hereditary and probably has no kinship with those already described. The bones are normal in all respects except that they are spattered with ovoid or round islands of dense bone (Fig. 382). It may be found at any age and causes no direct symptoms, although instances associated with other conditions which do cause symptoms, such as familial dermatofibrosis, have been recorded.

## CHAPTER XV

# DISEASES OF THE NERVOUS SYSTEM

## POLIOMYELITIS

(*Synonym*) Infantile Paralysis

POLIOMYELITIS is an acute disease in which there is widespread infection of the central nervous system. It is caused by a neurotropic virus of which there are three distinct strains all having a special affinity for the grey matter of the spinal cord. The basal ganglia and cortex are not as a rule affected severely, but occasionally they bear the brunt of the infection. The virus can be recovered from the central nervous system only after there is clinical evidence of infection, such as a rise in temperature. It is, however, present in the circulating blood some days earlier and this is now thought to be the route by which it reaches the central nervous system.

The changes in the spinal cord are of two kinds; firstly there is an inflammatory reaction around the capillary vessels of the grey matter and diffuse infiltration of the grey matter with polymorphonuclear leukocytes, and secondly there may be degenerative changes in the anterior horn cells. The affected cells may either be put out of action temporarily by pressure of the surrounding oedematous tissues, or they may be destroyed permanently by toxins produced by the action of the virus or as the result of deprivation of their blood supply.

### Epidemiology

The disease occurs in sporadic and epidemic forms. In Great Britain sporadic cases are reported every year in some numbers, mostly between the months of July and October; major epidemics have not yet occurred although from time to time sufficient cases are congregated in one locality to be regarded as a minor epidemic. The 1947 outbreak was of considerable size. In the United States of America the incidence is higher and severe epidemics are frequent. There is no doubt that the incidence is increasing throughout the world both in countries where it existed before and in those which, until recently, have been relatively immune. The arrival of the virus in a new country is often heralded, as has happened in Malta,

recognize its presence without animal inoculation, several extensive investigations have been carried out. It has been found that during an epidemic the virus can be recovered from the faeces of a large majority of the population, both children and adults, even though they have no clinical signs of infection. It is improbable that the virus is passed from person to person by direct contact because it is seldom that several members of one family, or those with medical care of patients, show clinical signs of the disease. Observations such as these have led some authorities to believe that during an epidemic infection is almost universal and therefore control must be impossible. On the other hand it has been found that there is a greater incidence of clinical infection amongst families containing a child of school age (although it is not necessarily the child that is affected) and this suggests that control should be possible. Attention is given to such problems as the contamination of food by careless handling or by flies, and contamination of water, both the main supply and swimming pools.

### Prognosis

The prognosis varies with different epidemics, and apparently with different strains of the virus. The mortality is greater amongst adults than children. The usual cause of death is respiratory paralysis, but it can be due to intercurrent disease, particularly in infants who may die of enteritis and marasmus.

There is complete recovery without any paralysis at any stage in a large percentage of those surviving the acute phase, the proportion varying from 25 per cent to 75 per cent in different epidemics. Recovery from paralysis depends on whether the anterior horn cells have been damaged temporarily or permanently; the extent of the initial paralysis, therefore, is not a reliable guide to prognosis, and some patients with severe initial paralysis return almost to normal.

### Clinical Features

Persons of either sex may be affected. The greatest incidence is usually from two to five years of age but it varies in different outbreaks and no age is immune.

There are three distinct clinical phases in the course of the disease:—

Phase of *onset*.

“ “ *recovery*.

“ “ *residual paralysis*.

**Phase of Onset.** The reaction to the initial infection varies greatly in different epidemics and in different individuals in the same epidemic. It may be:—

(b) *Abortive.* An influenza-like, pyrexial attack of brief duration, and unaccompanied by paralysis. Clinical diagnosis is presumptive and can only be made during an epidemic.

(c) *General Symptoms without Paralysis.* The general symptoms resemble those described below and there are identical changes in the cerebro-spinal fluid.

(d) *General Symptoms with Paralysis.*

(e) *Paralysis without General Symptoms.* The patient wakes in the morning unable to move certain muscles or limbs.

The incidence of sub-clinical and abortive infection is not known but it must be very high because of the frequency with which the virus can be recovered from the faeces during epidemics and the apparently high immunity of adults in many countries. It is probable that most of the population has been infected at some time or other without being aware of it.

**GENERAL SYMPTOMS.** In many instances the onset is marked by symptoms resembling those of any acute infection. The child is feverish, often for only a day, with a temperature of 100° to 101°, and there may be general malaise, headache, drowsiness, and perhaps diarrhoea and vomiting. Occasionally there is high fever and delirium. Muscular tenderness is usually a prominent feature and is evident early; pain on joint movement also occurs early, probably because this involves movement of muscles, and infants resent handling, nursing, and changing of napkins. At a later stage headache becomes more severe and there is usually pain and stiffness of the back and neck. Somnolence is sometimes marked, anorexia is common, and an initial diarrhoea frequently gives way to obstinate constipation.

Another mode of onset that is fairly often seen, at any rate in Great Britain, is characterized by more gradual development. There is an initial mild fever lasting two or three days, followed by a period of remission of a few days, and then a rather more severe, but indefinite, illness. Muscle pain and tenderness and stiffness of the neck may be delayed a little longer, and paralysis does not appear until perhaps a fortnight after the first symptoms.

The pressure of the cerebro-spinal fluid is raised and the cell count gradually rises to 50–250 per cubic millimetre. At first the increase in cells is shared by both polymorphs and lymphocytes, but the proportion of lymphocytes increases steadily. There is a gradual but moderate rise in both protein and globulin content of the C.S.F.

**PARALYSIS** often appears shortly after the general symptoms but it may be delayed for several days, and sometimes, when there has been no general disturbance, paralysis is the first feature to be noticed. Paralysis usually reaches a maximum within a few hours—gradual development is of bad prognosis. It is flaccid in type and may affect a single muscle, a group of muscles, or a complete limb or limbs. Sometimes there is weakness









known to increase the probability of paralysis and these must be avoided whenever practicable; the most important are fatigue and injury (and pregnancy). Paralysis often follows fatigue during the incubation period and it is the group or groups of muscles concerned that are likely to become paralysed. Thus the woman who uses a hand operated sewing machine during the febrile stage may suffer paralysis of the right upper limb, and the man who plays football may well develop quadriplegia. Some physicians regard the need for rest as being so important that they will not permit their patients to hold a book, or even feed themselves. Injury may also be followed by paralysis of the muscles of the same region; thus tonsillectomy may be followed by bulbar paralysis, and intramuscular injection by paralysis of the muscle injected. It has also been suggested that lumbar puncture is followed by an increased incidence of paralysis of the thigh muscles, but this is not proven.

After paralysis has developed rest is still the chief indication. Sedatives may be necessary for the relief of pain and to ensure rest. Aperients are required if constipation is troublesome but treatment should not be allowed to cause undue disturbance. The mattress must be firm, and if necessary supported by "fracture boards". Weak or paralysed limbs should be prevented from moving by sand-bags or light splints, the hips and knees being slightly flexed, the feet supported, and the shoulders abducted and in neutral rotation. At first rest is more important than accurate splinting and the latter can be postponed for a few days. Hot fomentations help to relieve muscle pain, but massage does more harm than good at this stage.

Paralysis of the respiratory muscles may require special attention. Atropine is given to diminish secretion. A mild degree of respiratory embarrassment can be alleviated by raising the head of the bed, but gross respiratory failure may develop very rapidly and the nurse should be warned that, if it does, artificial respiration must be commenced manually and continued without interruption whilst the patient is being transferred to a mechanical respirator.

**Phase of Recovery.** More active treatment is begun when the general symptoms have subsided but first it is essential to record accurately the precise extent of muscle weakness and paralysis. This is of importance in determining the prognosis and in deciding the earliest moment at which operative interference is desirable. It is not sufficient just to note, say, "foot-drop," but each muscle must be examined separately. The method of recording muscle power has been standardized in Great Britain by the Medical Research Council and this should always be used (p. 506). Progress must be charted at regular intervals.

**SUPPORT.** This is necessary to prevent deformity and to rest weak or paralysed muscles. The general requirements are that the support should be so arranged that paralysed muscles are held at an intermediate point in

The exact nature of the support varies with the extent of the paralysis. If it is widespread and includes the muscles of the buttocks, back or abdomen, a plaster bed is very satisfactory. It has the advantage of being able to accommodate all four limbs, it is comfortable, and it allows active movements of unparalysed muscles (Fig. 383). A single extremity without involvement of hip or shoulder is conveniently supported on a plaster shell.

The position in plaster plays an important part in determining subsequent function and must be arranged with meticulous care. The spine requires the same curves as when standing; the hips and knees should be slightly flexed, the ankles at right angles with the legs and the feet plantigrade, neither inverted or everted; the arms should be abducted about 70° in the scapular plane and in neutral rotation; the elbow is at a right angle, the wrist is slightly dorsiflexed, the fingers are placed in the position of rest and the thumbs partly opposed. The arm of the child illustrated below is abducted too much.

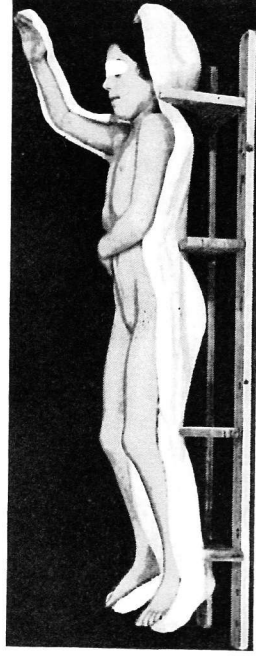


Fig. 383.

**MOVEMENT.** Commencing as early as possible, every joint should be gently moved once daily through its greatest painless range. This is an important feature of early treatment and has the object of preventing joints from becoming stiff and muscles from deteriorating.

Active contraction of affected muscles is encouraged but excessive fatigue must be avoided. At first a muscle may give a flicker once or twice but repetition is impossible until after a long rest; later it may move the joint whilst the weight of the limb is supported, and later still move it against gravity. Whatever the power of the muscle, exercise must be arranged to suit its strength, and it must be followed by a period of rest long enough to allow it to recover from fatigue. Trick movements should be watched for and care taken to encourage use of the proper muscles instead. Immersion in water during exercise is of great value because it counteracts the effect of gravity.

Co-ordination of movement and "awareness" of certain muscles appear to be lost in some patients, perhaps because of damage to communicating fibres within the spinal cord. Deliberate exercises in co-ordination may,

PASSIVE PHYSIOTHERAPY is of assistance in maintaining the nutrition of very weak or paralysed muscles. Muscles function better when warm than cold and therefore they may be gently heated before exercise. Massage improves both venous and lymphatic circulation but it should not be carried out roughly, nor for too long, or it may damage muscle fibres. Electrical stimulation may also be helpful. A paralysed muscle loses its response to faradism but reacts to galvanism and therefore interrupted galvanic stimulation is a useful form of exercise.

PROGRESS. No precise rules can be given as to the time when sitting up or walking may begin because of the great individual variation in the severity and distribution of paralysis. Every muscle with power less than that necessary to contract against resistance requires support until maximum recovery has occurred; this is difficult to arrange in an ambulant

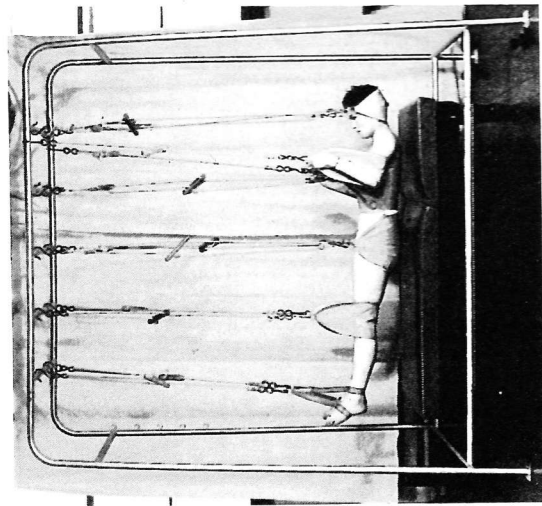


FIG. 384. Guthrie-Smith suspension frame.

patient, except when paralysis is limited in extent, and therefore many patients have to remain recumbent for several months. When only one lower limb is affected, however, walking may be possible fairly early using crutches and a light plaster splint; the upper limbs can be supported on splints or by means of a plaster spica; and the abdominal muscles must be protected by a well-fitting cloth corset if they show any sign of weakness.

Support for the trunk is very important when the erector spinae are paralysed, especially in children, in order to prevent the development of scoliosis. There is no really efficient splint or jacket and therefore recum-

place; this is often as long as two years and sometimes even longer. Exercise is carefully graduated to develop the weaker muscles. A swimming pool, or if this is not available, suspension by springs or by weights and pulleys, are of value when exercising weak muscles because the force of gravity is reduced (Fig. 384). It is most important to plan exercises with special regard to muscle balance because, if the patient is left to himself, he naturally uses the stronger muscles which then develop at the expense of the weaker ones, and imbalance is increased.

The final result, provided the destruction of nervous tissue is not too great, depends to no small extent on the perseverance of the patient. The way in which some apparently hopelessly paralysed people make use of the little power they have is so remarkable that it should never be forgotten by those in charge of them. The patient must not be allowed to despair however dreadful his disability may seem, but he must be treated in an atmosphere of encouragement and cheer, guided through the difficult early days, and taught to understand the way in which he can best help himself.

### Deformity

Deformity can be caused by (1) overaction of unopposed muscles, (2) relative shortening of paralysed muscles and (3) weight-bearing on a partly paralysed limb. It should, however, seldom occur when treatment is carried out in proper conditions and is competently supervised, except perhaps at the spine which is difficult to support by mechanical means.

Some deformities, such as genu recurvatum, are associated with lengthening of the soft tissues and the joint is hypermobile. In the majority, however, the deformity is fixed by soft tissue contraction, and at a later stage by adaptive changes in the shape of the bones.

**Spine.** Scoliosis is a common sequel to weakness or paralysis of the muscles of the back or abdomen, particularly when unilateral. In children, the deformity tends to increase during growth and may become very severe. It is probable that "idiopathic" scoliosis is often due to undiagnosed poliomyelitis (p. 109).

Scoliosis is often rather late in developing and is easily overlooked in the early stages. Once the shape of the bones has changed it is impossible to correct it; prophylaxis is therefore most important and a spinal or abdominal support is necessary whenever the muscles of the trunk are involved. The support must be worn until growth of the spine has ceased at about fourteen years of age, and then spinal fusion may be considered. Developed scoliosis is treated in the same way as when it is due to other causes (p. 115).

**Hip.** Flexion-adduction deformity occasionally results from paralysis of the gluteus maximus, and when all the glutei are paralysed the hip may dislocate backwards. A mild degree of flexion deformity can be overcome by stretching, but a more severe contraction may require correction by

may be controlled by construction of an artificial check ligament (*Ober*), and several other operations.

**Knee.** Various deformities may occur. Flexion deformity due to overaction of the hamstrings and gastrocnemii can usually be straightened by traction or by a wedged plaster. Hyperextension due to weakness of the flexors is a serious disability; it is not satisfactorily treated by operation and a caliper may be necessary. Genu valgum is not uncommon, and if severe it may need correction by osteotomy (p. 31).

**Foot and Ankle.** Pes equinus, probably the commonest of all paralytic deformities, is due to paralysis or weakness of the dorsi-flexors of the foot and relative overaction of the calf muscles. The deformity is easily corrected by lengthening the tendo-Achillis but this must not be done if the knee is unstable. Many patients whose quadriceps are paralysed can walk without apparatus by bracing the leg against an equinus foot.

A cavus deformity develops when the triceps suræ are paralysed and the tibialis anterior together with the long flexors and the invertors or evertors of the foot remain in action. The severe degrees are most difficult to correct, even at operation, and every effort should be made to prevent them. There is some evidence that they may be at least reduced in severity by tendon transfers designed to provide a better balance of the muscle power available. Stabilization should, however, be performed as soon as practicable.

A valgus or varus deformity occurs either alone or in conjunction with equinus or cavus as a result of an unequal degree of weakness of the invertors and evertors. A valgus position is also assumed by a flail foot when bearing weight. Tendon transfers have only a limited application as an adjunct to other operations, and occasionally to restore the balance of the foot when only a single muscle remains paralysed.

**Upper Limb.** Fixed deformities, except at the shoulder, are seldom seen and they do not conform to standard types. Adduction-internal rotation deformity of the shoulder is sometimes troublesome and may be overcome by repeated passive stretching or by operation (p. 526).

### Residual Paralysis

The assessment of a patient at this stage is a responsibility needing a wide knowledge both of the mechanics of the human body and of the many different kinds of stabilizing operations and apparatus. Maximum motor recovery has taken place, or at any rate the possibilities of further improvement are defined, and the problem is to enable the patient to make the greatest possible use of the remaining muscle power.

It is not always necessary to await maximum recovery before embarking on stabilizing operations provided the patient is old enough for arthrodesis to be practicable. A muscle that shows no recovery or only a flicker of movement after, say, six months is unlikely ever to become strong

the better because the immediate improvement in function enables the patient to concentrate on other and more difficult problems. But if the muscles of the hand are weak, the decision to operate should be postponed for as long as there is hope of improvement.

Three methods of stabilizing joints and redistributing muscle power are available for use separately or in combination—*apparatus*, *arthrodesis* and *tendon transplantation*. Much ingenuity has been displayed in evolving different uses for tendon transplantation, and although they are of great value for an occasional patient, few have proved successful enough for general service. In Great Britain the use of transplants is more or less

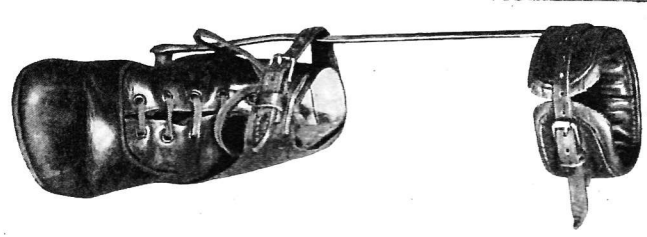


FIG. 385. Short inside iron and outside I-strap to control pes varus.

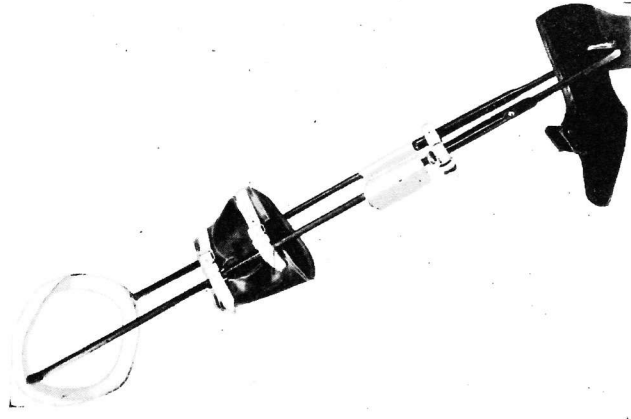


FIG. 386. Long double irons with round socket in heel and ankle stop at 90°.

restricted to the hand; for the lower limb arthrodesis is more popular because it usually gives a better functional result.

Detailed description of the many possible procedures is outside the scope of this book and only the general lines of some of the commoner ones will be indicated.

**Spine.** The approach, whether by apparatus or operation, is similar to that for scoliosis in general but the paralytic spine is more easily supported by apparatus (p. 115).

**Hips, Knees and Feet.** *Bilateral.* Almost every patient whose arms



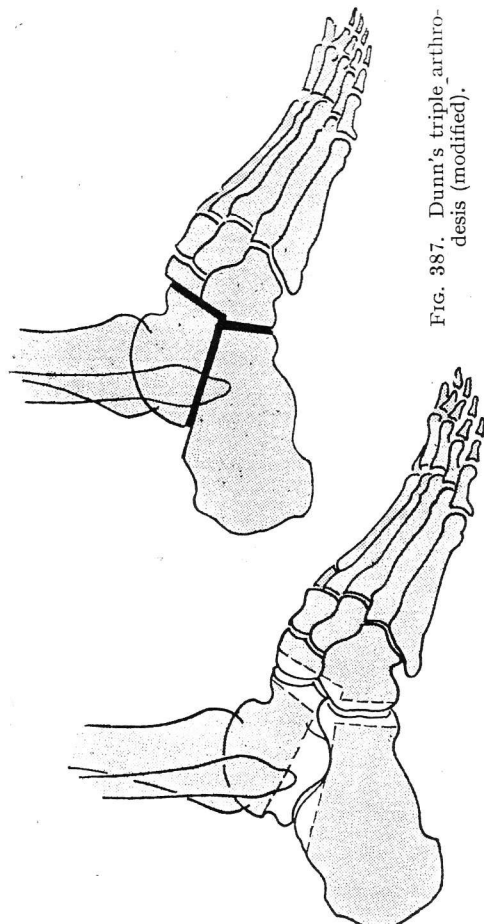


Fig. 387. Dunn's triple arthrodesis (modified).

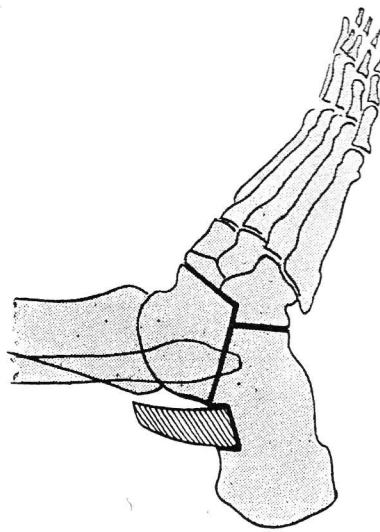


Fig. 388. Campbell's bone block for foot drop.

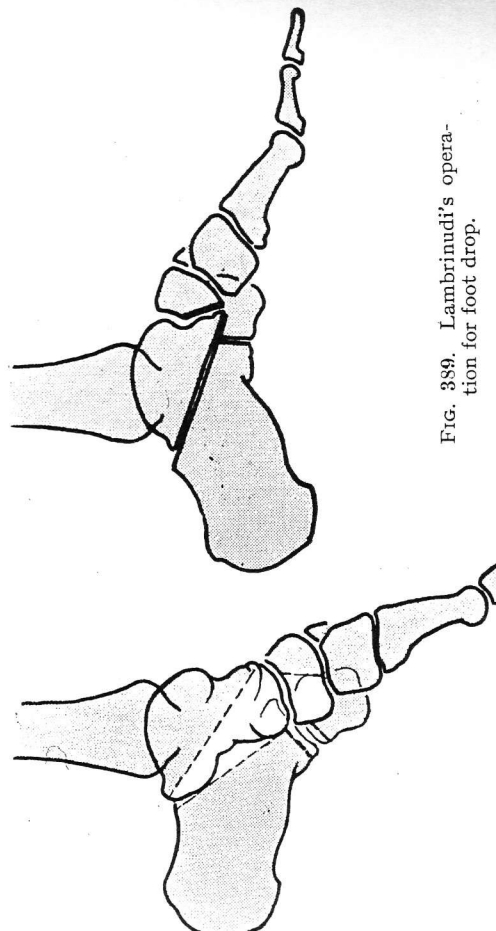


Fig. 389. Lambrinudi's operation for foot drop.

spinal muscles also are weak, a spinal support is provided, and then "tripod" walking is possible with the aid of two crutches. Progression by this method is slow, but at least it is possible for the patient to get about the house alone. Stabilizing operations are seldom worth while.

**Unilateral.** Absence of the glutei causes a gross "Trendelenberg limp" which is very difficult to prevent (p. 144). Leg irons can be attached to a pelvic band or spinal support by means of a hinge at the hip, but this is seldom satisfactory. Arthrodesis of the hip does not give good function unless the quadriceps are strong. As a rule, when the knee and ankle are flail as well as the hip, it is best to accept the limp and control the knee and foot with irons. Occasionally *Ober's operation* may be feasible; the fascia femoris is detached at the lower end, passed through a hole cut in the upper end of the femur and attached to the erector spinae which are thus enabled to act as abductors of the hip.

**Knee and Foot.** The gait, if only one leg is affected, is reasonably good when the knee and foot are controlled by irons. It is often possible, however, to dispense with apparatus, even when both legs are involved, by stabilizing the foot in equinus (see below).

**Knee.** Paralysis of the quadriceps causes surprisingly little disability provided there is some fixed equinus against which the leg can be braced. The glutei, when reasonably strong, are used to brace back the thigh thus fixing the knee, and there is hardly any limp; when the glutei are weak, the trunk is thrown a little forwards when walking and then the knee is forced backwards and locked because the centre of gravity of the body is in front of the joint (Fig. 4, p. 7). Although it is unusual for apparatus to be necessary when only the knees are affected—I have a patient with bilateral flail knees and stabilized feet who dances excellently—sufficient stability cannot always be acquired without assistance and then leg irons with a knee hinge and lock can be supplied. With unilateral paralysis arthrodesis of the knee is sometimes desirable. Transfer of the hamstrings to the extensor mechanism has often been tried but it has seldom proved to be worth while.

Paralysis of the hamstrings in the presence of good quadriceps is difficult to deal with and often necessitates the use of apparatus.

**Foot.** Stabilization of the foot is a really satisfactory operation giving good function and practically no limp. It is indicated when there is paralysis of the invertors causing a valgus foot, paralysis of the evertors causing a varus foot, and when the foot and ankle are flail.

The several operations in common use are all based on the classical prototypes described by Dunn in England and Hoke in the U.S.A. *Dunn's operation* is a triple arthrodesis in which the talo-calcaneal and calcaneocuboid joints are fused and the entire navicular is removed, the talus being joined to the cuneiforms. Most surgeons prefer not to remove the navicular and fuse the talo-navicular joint instead (Fig. 387). The same object may be achieved by a triple arthrodesis in which the talo-calcaneal and calcaneocuboid joints are fused and the entire navicular is removed, the talus being joined to the cuneiforms. Most surgeons prefer not to remove the navicular and fuse the talo-navicular joint instead (Fig. 387). The same object

way as to limit foot drop and yet permit dorsi-flexion at the ankle. The desirable range of movements depends on the height of heel customarily worn and on whether the knee is flail; in the latter case some fixed equinus must be provided. In *Lambrinudi's operation* a wedge of bone of the required size is excised from the talus and calcaneus and this enables the necessary range of movements to be provided with accuracy (Fig. 389.) Other procedures having the same objective, such as *Campbell's operation*, involve first a triple arthrodesis, and then checking foot drop by a bone graft inserted into the posterior end of the calcaneus (Fig. 388).

**Upper Limb.** The value of the upper limb depends principally on the function of the hand, and as long as movement of one finger remains, the hand can be made of some use. It is, however, necessary for the hand to be moved to the object to be grasped and therefore, if the shoulder and forearm are flail, they must be stabilized.

**Shoulder.** Arthrodesis is a most useful operation when the muscles controlling the scapula are reasonably strong (p. 338).

**Elbow.** Transfer of the lower fasciculi of the pectoralis major to replace the flexors of the elbow (*Clark's operation*) is a valuable procedure and can give really useful power.

Stabilization may be effected either by arthrodesis or by an arm-corset with a hinge that enables the joint to be locked in any position (Fig. 265, p. 353). Arthrodesis is usually more satisfactory when the muscles of the shoulder are strong enough to control it, but a corset is better when the shoulder has been arthrodesed.

**Hand.** The possibilities depend on the available muscle power. The essential requirements are active flexion of at least one finger, preferably the forefinger, and a "post" against which the finger can press. If all the flexors of the fingers are paralysed, it may be possible to supply motor power by transplanting one of the flexors of the wrist. The best "post" is the opposable thumb, and if a suitable muscle is available, a tendon transplant can be performed to provide active opposition; if no muscle is available, the thumb may be fixed in opposition by means of a bone graft secured between the first and second metacarpals. It may be desirable to arthrodesis the wrist in slight dorsi-flexion (p. 379). When paralysis is limited to the intrinsic muscles and there is a claw hand, *Bunnell's operation* is successful (p. 377).

### Resettlement

Sufferers from poliomyelitis are remarkable for the fortitude with which they face their problems and overcome their physical disabilities. It is rare for residual paralysis to be so widespread as to leave the individual quite helpless. Most people, by learning to do the ordinary things of life in a different way from others, are able to look after themselves and

## SPASTIC PARALYSIS

### SPASTIC PARALYSIS

(*Synonyms*) Little's Disease, Cerebral Paralysis of Children, Cerebral Diplegia, Congenital Diplegia

Paralysis is a misnomer. The term "spastic paralysis" is applied to a heterogeneous group in which there is disturbance of the locomotor system with hypertonia of the muscles, occasionally hypotonia, often athetosis, but never paralysis. The condition is present at or soon after birth and is due to a lesion of the cerebral cortex or basal ganglia causing a disorder of upper motor neurone type. The precise symptoms vary with the extent and distribution of the lesions and include spasticity, involuntary movements and mental defects.

Different names are applied according to the region affected:—

- One extremity. *Monoplegia.*
- Half the body. *Hemiplegia.*
- Both legs. *Paraplegia.*
- Both legs and arms. *Diplegia* or *quadriplegia.*

The etiology has been much disputed. Of the several theories advanced at different times, the following are usually accepted:—

**ANTE-NATAL**—Rare. Early arrest in foetal development.

**NATAL**—Common. Birth injury causing meningeal hæmorrhage is probably the cause in most instances.

**POST-NATAL**—Uncommon. Thrombosis, meningitis, or encephalitis.

### Clinical Features

The symptoms vary with the distribution of the degenerative changes in the brain. They fall into four distinct groups which, however, are often mixed.

**PRE-FRONTAL CONVOLUTIONS.** Mental defects which vary in degree from slight backwardness to idiocy.

**PRE-CENTRAL CONVOLUTIONS.** Defects of motility. There is spasticity of the lower limbs and often of the upper limbs also. Very occasionally there is hypotonia affecting the limbs, the back and the neck.

**BASAL GANGLIA.** Uncontrolled athetoid movements of the affected limbs, usually more marked in the arms than the legs. Athetosis is exaggerated on attempting voluntary movements and during emotional stress, and it is usually absent during sleep.

**CEREBELLUM.** Rare. Ataxy of cerebellar type.

Phelps has drawn attention to a condition of pseudo-spasticity, or "athetosis with tension," which occurs as the result of spontaneous efforts to control involuntary movements. The limbs are held rigid in the attempt