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# The surgical management of cerebral palsy

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Cerebral palsy more than any other paralytic condition presents some of the most difficult and challenging problems in orthopaedic surgery. It is only relatively recently that surgery has come to play an important role in the treatment of this condition, but sufficient experience has now been gained to make possible an overall appraisal and statement of its value and indications.

Cerebral palsy is a complex and heterogeneous mixture of syndromes, but it can be conveniently defined as "a permanent motor disorder appearing before the age of three years due to a non-progressive interference with development of the brain". (MacKeith *et al.*, 1959).

In other words (Holt, 1965):

(1) It affects young children.

(2) It persists throughout life.

(3) There is always some disorder of muscle function.

(4) In all cases there has been interference with the developing nervous system.

The various manifestations of the condition combine with one another in many different ways, making assessment difficult and time consuming. The more important features are:

Ataxia—meaning inco-ordination and poor balance.

Athetosis—the occurrence of bizarre, purposeless movements.

Hypertonia or hypotonia—alterations of muscle tension.

Spasticity—meaning hyperactivity of the muscle stretch reflex.

Rigidity—persistent stiffness of muscles and movements.

Tremor—meaning rhythmical purposeless movements.

The disorder characteristically affects groups of muscles. It may affect a single limb, when it is described as monoplegia; arm and leg on one side, or hemiplegia; both legs—paraplegia; or all four limbs—quadriplegia. The term diplegia is used to refer to the latter condition when the paralysis of the legs predominates. In practice, however, in the majority of cases, careful examination will reveal abnormalities in limbs which at first sight appear to be normal. The motor problems are frequently complicated by other handicaps, such as defects of communication, blindness, and commonly mental abnormality, which in many cases is severe. This inevitably makes management a complex problem and a team approach is usually advisable. The team basically consists of a paediatrician, a mental health expert, a physiotherapist, and an orthopaedic surgeon. Such a team can attempt to build up a complete picture of mentality, specific handicaps, orthopaedic problems and social difficulties, and all these factors can be considered in making treatment decisions.

Although the contributions of orthopaedic surgery have been considerable, the actual operative techniques involved are for the most part relatively simple; nevertheless considerable judgement is necessary to derive the greatest benefit from them.

The basic aims can be defined in the simplest terms as being:

(1) To prevent the occurrence of serious deformity.

(2) To utilize to the best effect the resources which the child possesses.

It is in the first of these aims that the orthopaedic surgeon is likely to play the greatest part. In making orthopaedic decisions information will be required from the rest of the team about the child's overall capacity for function and the possibilities for rehabilitation and training. At best, however, these can only be rough guides and in practice it often pays dividends to aim high rather than low, because surprisingly often these children can achieve a remarkable level of function and independence

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when at first sight the situation might seem hopeless. The orthopaedic surgeon of course will himself be responsible for a large part of the functional assessment.

**Overall function**

This always starts with an assessment of the child's capabilities at the time of examination; for example, the ability to control head and limbs, to crawl, stand or walk, the ability to manipulate objects and to co-operate, and the presence of abnormal movement patterns, joint instability and so on. This necessitates a careful clinical record, wherever possible supplemented by photographs, and in some cases by cinephotography.

**Deformities.**

The measurement of deformities is carried out in a systematic manner, taking each joint in turn, not forgetting the spine. It is important at this stage to make a distinction wherever possible between fixed deformity and mobile deformity. Fixed deformities are those where it is not possible to put the joint into the anatomical position. Mobile deformities are those where the joint is continuously, or for most of the time, held in the deformed position, but can in fact, given sufficient relaxation, be placed in the anatomical position. It will frequently be possible to make a judgement about the relative importance on the one hand of soft tissue contractures, involving capsules, ligaments, tendons, and sometimes skin; and on the other hand the bony deformities which usually develop in longstanding cases. Both of these types of fixed deformity may arise together with, or independent of, spasticity. The distinction between spasticity and true joint contracture is of the greatest practical importance. Even when moving the joint gently, which should always be the case when examining these patients, it may not always be possible to estimate the amount of true contracture, and on occasion a decision about a corrective procedure may have to be made on the basis of an examination under anaesthesia.

There are a number of common patterns of deformity and it is therefore to some extent possible to standardize the indications for surgery.

**Specific motor and sensory function.**

Abnormalities of muscle function are of course the fundamental problems of cerebral palsy. Various defects of function are possible, indeed the phenomenon of muscle contraction and muscle integration can be disturbed in many ways. To give a few examples—the onset of a muscle contraction in relation to its stimulus may be delayed abnormally and the strength of contraction may increase relatively slowly. Or again the muscle may fail to relax at the correct time, antagonists may be out of phase with prime movers, and general muscle integration may be absent even though individual muscles may be capable of almost normal function. Sometimes stimulation causes too wide a response, and again, some children lack sensory awareness of muscle action, so obviously find difficulty in learning skills. It is important to consider the power of contraction. There is naturally a tendency to concentrate on spasticity and tightness of muscles, but careful examination will frequently reveal differences in relative power of muscle groups (Tachdjian and Minear, 1956), and occasionally weakness may be the sole sign of cerebral palsy. It seems likely that weakness is an important factor in the development of deformities as in other paralytic conditions (Sharrard, 1961).

Sometimes a muscle group is consistently weak but occasionally only certain movements are involved, so that a muscle may appear weak in one movement and not in another. I have already mentioned the difficulty of co-ordination in some patients, and of course there is often the problem of the child not understanding what is required. Frequently much time is necessary to gain the necessary co-operation. Given this co-operation, it is usually possible to arrive at some measure of muscle power, and indeed, it may sometimes be possible to use the Medical Research Council scale of grading. The situation, however, is much more complicated than in other paralytic conditions, and to see the problem as a simple one of rebalancing weak and strong muscles, is to invite many failures in treatment. The difficulty is that, although relative weakness may be demonstrable, the muscle or muscle group which appears to be weak may be capable of powerful contraction given the appropriate conditions. It is not uncommon to find that when the strong muscle group has been weakened by tendon section or denervation, the anta-

gonists develop a totally unexpected strength which may result in deformity occurring in the opposite direction. This suggests that much of the weakness was in fact inhibition from the strongly acting antagonists and it is therefore unwise to deduce too much from the apparent discrepancies in power. Undoubtedly spasticity in a group of muscles has this effect of inhibiting the antagonists and producing an apparent or indeed a real power discrepancy, with subsequent development of deformity. Theoretically the best treatment in these circumstances would be to diminish the spasticity whilst leaving the power intact. This would, however, require subtle interference with neurology and is hardly possible at the present time, although phenol nerve injection techniques suggest that such a differential block can be achieved in practice. (Braun *et al.*, 1972).

Other factors also come into play in producing relative muscle weakness. A stretched muscle may be weak because of mechanical disadvantage and if this disadvantage is rectified power may again be greater than expected. In practice of course an assessment of muscle power is of value in that it gives some idea of the speed at which deformities are likely to progress. The greater the difference in power, the more rapidly will the deformity occur, but even slight weakness can cause severe disability from simple functional loss long before the development of severe deformity. For example, relative weakness of the hip extensors or wrist extensors can produce severe interference with gait or hand function, even though deformity may develop relatively slowly.

Spasticity or hyperactivity of the stretch reflex is a difficult phenomenon to measure. It varies in degree and a simple clinical assessment can be made by moving the joint rapidly and noting where the 'catch' occurs. The presence of clonus may be a valuable physical sign in picking up milder degrees of spasticity. Again of course this is a manifestation of the hyperactive stretch reflex.

Co-ordination problems such as ataxia and athetosis are important defects of motor function and their assessment is important, although they are not usually amenable to surgical correction, and are not in themselves

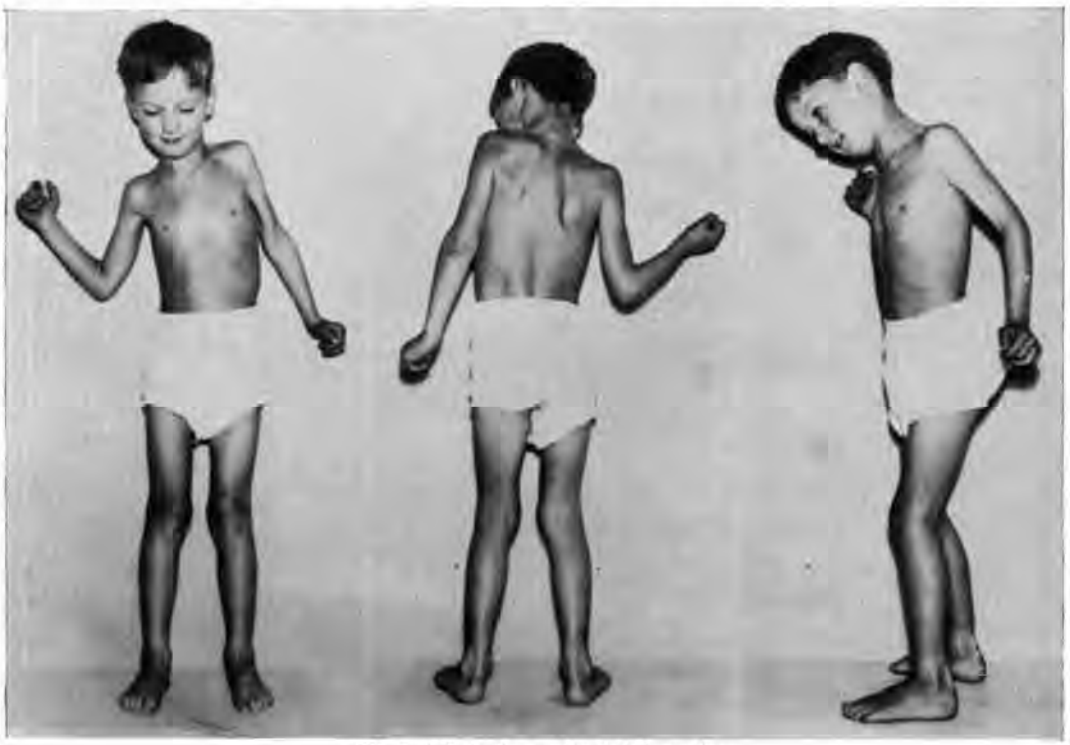

Fig. 1. Athetosis—no fixed deformities.

responsible for the development of deformity (Figure 1).

I have already mentioned the aims of surgery which are to prevent or correct deformity and to utilize resources to best functional effect. In practice these two aims tend to be complementary, in that procedures which are utilized to prevent deformity will also produce improvement in function. In deciding whether surgery is necessary, the assessment will normally have drawn attention to two main factors. Relative *muscle imbalance* usually associated with spasticity on the strong side, and either already associated with *deformity* or suggesting its potential development. It is rarely possible to detect potential deformity before it has already begun to occur, but the aim should always be to anticipate developing deformity and to deal with it before it becomes severe. Accepting that a deformity is present, the first question will usually be—*how can the deformity be corrected*?

Very mild deformities can sometimes be corrected by simple physiotherapeutic procedures such as stretching and serial plasters. More severe deformities will require elongation of tendons, capsules and ligaments, and sometimes skin. As in other paralytic conditions the more severe degrees of deformity in the older child may require bony surgery to produce adequate correction. Better results are always achieved by surgery at the soft tissue stage and, provided this is performed sufficiently early and adequately, it is usually possible to avoid the development of the worst bony deformities.

Having decided on the method of correction, the next question to be answered will be how to overcome the relative muscle imbalance. As already mentioned this is difficult to achieve because of the problems of spasticity and inhibition and the uncertain effects of mechanical disadvantage. In practice the methods which seem to work best usually combine well with the soft tissue correction procedures. For example, lengthening of the tendo-achilles at the ankle will often correct deformity and also will reduce the relative over-action of the calf. Whether it does this by simple reduction of mechanical power or by diminishing the apparent stretch inflow and therefore the corresponding inhibition of the ankle dorsiflexors is somewhat difficult to assess. The important point is that it does work and tendon elongation or even simple tendon section is a very valuable procedure in the surgery of cerebral palsy. Similarly, partial denervation of a muscle group may produce its effects in several different ways, but again experience shows that there are many situations where it can be conveniently and safely utilized to produce a re-balancing effect.

Because of the problem of assessing true muscle power the tendon transplantations which are useful in other paralytic conditions are on the whole less valuable in cerebral palsy. They are extremely variable in their effects, the commonest problem being overcompensation resulting in deformity in the opposite direction, and one can imagine that in disorders of stretch reflex arcs the effects of tendon transposition must be complex in the extreme. The situation is somewhat akin to the difficulties of tendon transplantation of reflexly innervated muscles in spina bifida. There are, however, a few situations where transplantation appears to be valuable—in deformities of the wrist particularly, and in certain deformities of the foot. In practice the most widely used re-balancing procedures are tendon lengthening and partial denervation.

Having decided, then, on the best method of achieving a balance, the next problem to be considered will, as usual in paralytic conditions, be how to achieve joint stability. Fortunately in cerebral palsy this is usually much less of a problem than in flaccid conditions, because although there may be true paralysis, there is usually a good deal of active musculature around the joints, and the corrective procedures and re-balancing operations are frequently sufficient to provide adequate joint stability.

Although splints and braces undoubtedly play a part in the overall management, it is often possible to avoid their use by judicious surgery. Modifications to footwear of course are frequently necessary but calipers are seldom needed. Again, as in other paralytic conditions, arthrodesis plays little part. One of the few widely used fusion procedures is that of triple arthrodesis in the older child to achieve a final stabilization of the foot. Arthrodesis of the wrist has been popular, but is losing some of this popularity in favour of re-balancing operations such as muscle slides and tendon transplantation.

Finally the question arises of restoring function in individual movements. Again the corrective procedures will often have the effect of achieving this end. Occasionally a specific procedure will be of value. For example, a particular hand or finger movement may be restored by an appropriate tendon transplantation. Opportunities for this are uncommon, however.

It is convenient next to consider the problems which arise with individual joints and then to discuss some of the patterns of deformity which occur and the way in which the various procedures can be utilized.

Deformities of the spine are fortunately rather uncommon in cerebral palsy, but all of the usual deformities do occur and can be formidably severe. Scoliosis may be associated with lordosis or kyphosis and one also occasionally sees curious torsional deformities, where the whole child appears to have been twisted round the vertebral axis, with torsion of the spine and pelvis, and the hips deformed in such a way that one is adducted and the other abducted. At the present time most of these spinal problems are nowhere near a solution, although the standard techniques for dealing with scoliosis are being utilized. There is usually no difficulty with sensation in these patients, so that external splintage may be used, and the Milwaukee brace and corrective spinal plasters and supports all have a place. The same applies to the tried and tested surgical techniques of fusion and internal fixation. Nevertheless the deforming forces are very difficult to overcome and it seems likely that until the exact forces involved can be better understood and rectified, this problem will remain a very difficult one.

The hip joint is often the site of trouble in cerebral palsy, the commonest deformities being flexion and adduction, together with internal rotation (Figure 2), although occasionally other patterns are seen—abduction and

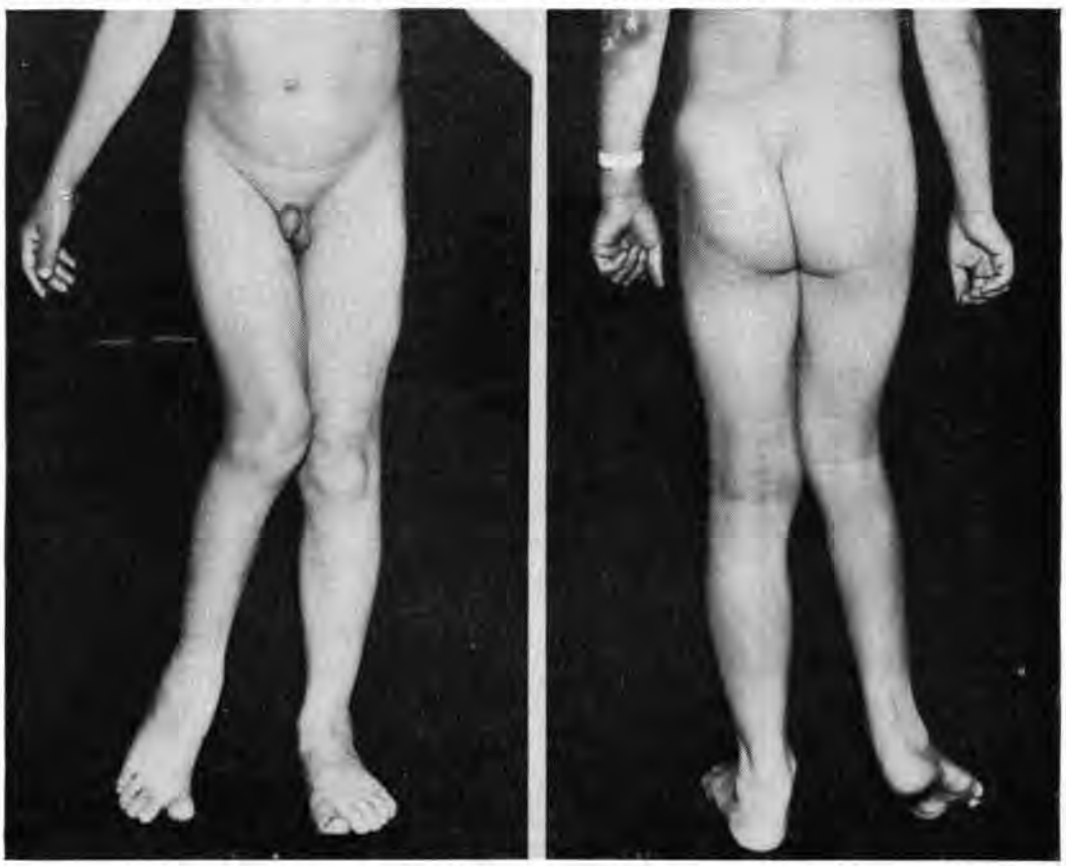

Fig. 2. Spastic hemiplegia.

external rotation being much less common. One of the commonest problems in cerebral palsy is a gradual diminution in the abduction range of the hip, and sometimes also in the range of extension, associated with tightness and usually spasticity of the abductors and flexors. This may, if left untreated, lead to gradual subluxation of the hip with eventual dislocation (Tachdjian and Minear, 1956, Sharrard *et al.*, 1975) and considerable increase in the disability (Figure 3). Careful and frequent orthopaedic examinations will detect this diminution in range and relatively simple surgical intervention will usually prevent it. It should be remembered that subluxation may occur before the abduction range diminishes to nil and is often associated with abnormalities in the shape of the femoral neck and its relationship to the shaft.

Adductor tenotomy is a simple and valuable procedure and it is a convenient guide to say that when abduction range has diminished to 30 degrees then surgery will normally be necessary. The adductor tenotomy is best performed by an open technique and consists of division of all tight structures, and where spasticity is severe, a section of the anterior branch of the obturator nerve. It is of the greatest importance to avoid dividing both branches of the nerve, as the disability from this is very severe. A short period of splintage in abduction after the operation is then usually followed by a period of physiotherapy to restore the original level of function.

At this point it is worth mentioning that in cerebral palsy the older the child, the more a surgical procedure temporarily interferes with function, and this may be a great disappointment to the parents who often feel that the child has been made worse by the procedures. This is particularly the case with hip surgery and it is therefore important to explain this point beforehand.

Occasionally section of the flexors, particularly the psoas, will be necessary to prevent progressive flexion deformity. Again if performed early and repeated this will usually avoid the development of the very severe flexion deformities seen in the untreated child. The dislocated spastic hip is a very difficult

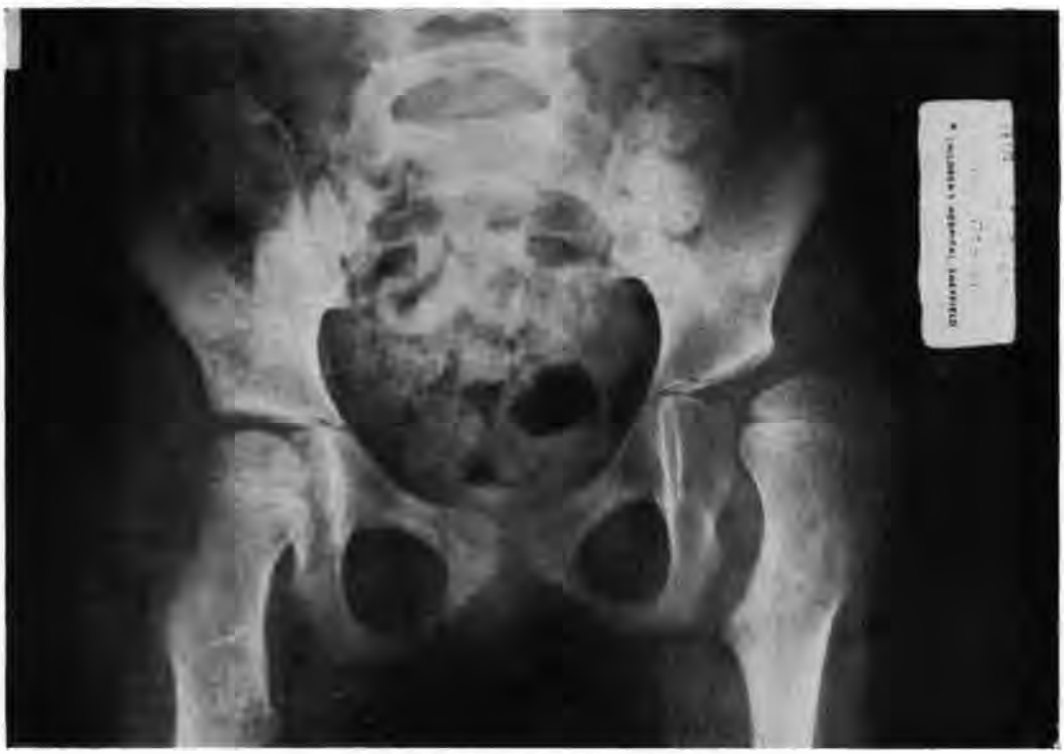

Fig. 3. Subluxating hip in cerebral palsy.

problem to deal with, and before embarking on the extensive surgery which is usually required to replace it, it is necessary to decide how much the dislocation is contributing to the overall disability and how likely one is to achieve permanent stability.

Rotational problems of the hip may be part of an overall pattern of deformity and are sometimes improved by more distal procedures. In persistent cases rotational osteotomy is usually a safe and simple procedure, but again in the older child its benefits must be set against the drawback of the considerable setback in function which usually occurs.

Although walking aids may be necessary to maintain the upright posture and to provide balance and stability it is rarely necessary to provide bracing to control the hips.

At the knees the commonest deformity is a flexion contracture, again usually associated with spasticity of the hamstrings and often forming part of a characteristic pattern of deformity of the limb (Figure 4). Less commonly hyperextension of the knee occurs, as occasionally does valgus or varus angulation. It should be possible to anticipate deformity and to carry out the necessary surgical procedures at an early stage. Hamstring elongation, or occasionally excision, is the most useful procedure (Figure 5). One of the difficulties about this procedure is the very strong tendency for the hamstrings to reform, or, even when they have been transplanted, to find their way back to their original insertions. Eggers' operation (Eggers, 1952), which is a transplant of the hamstrings into the lower femur, was designed to remove the flexion force at the knee and also to provide extra extension force at the hip. Its benefit however appears to stem mainly from the removal of the flexion force and its effects

Fig. 4. Flexion contracture of knees.

Fig. 5. Hamstring release for flexed knees.

as an active transfer have probably been overstated. It does have the slight advantage of making re-attachment of the tendons less likely.

If there is flexion contracture of the knee it is usual to elongate the distal hamstring tendons. It is convenient at this point to mention the curious condition of proximal hamstring tightness associated with difficulty in straight leg raising and in achieving an adequate length of stride, resulting in a rotating pelvis type of gait. This can be conveniently dealt with by proximal hamstring section (Sharrard and Seymour, 1968), a valuable procedure which may need to be repeated several times during the total period of growth.

At the ankle and foot a wide range of deformities are possible. The commonest by far is equinus deformity of the ankle associated with tightness of the tendo-achilles. Also common are cavus deformities of the foot with clawing of the toes and the valgus foot with a prominent head of talus on the medial side of the foot.

As already mentioned the various joint deformities tend to be associated together in patterns. The commonest one and the one which is typically associated with cerebral palsy of the hemiplegic type is that in which most of the joints are flexed; the elbow and wrist being flexed, fingers flexed into the palm, hip flexed, adducted and internally rotated, the knee flexed and the ankle in equinus. This pattern, more than any other, exemplifies the importance of tackling the problem step by step rather than embarking on an ambitious programme of surgery, much of which may not be necessary. It may be found, for example, that a simple elongation of the tendo-achilles will be enough to allow the heel to reach the ground and will remove much of the stretch stimulus from the calf muscles resulting in a general diminution of spasticity over the whole leg, and even occasionally in the arm. As a result the knee flexion and the hip flexion become less, and the internal rotation at the hip may diminish. In other words, one simple distal procedure has achieved at least partial correction of all the deformities. Having done this and allowed the child to recover it can then be decided if further procedures are necessary, usually proceeding from distal ones to more proximal.

Other patterns are also common. The scissoring of the diplegic due to adductor spasm and usually associated also with equinus of the ankles is very characteristic, (Figure 6) and can lead to great difficulties, even when sitting in a chair.

Fig. 6. Scissoring in spastic diplegia.

Also commonly seen in the diplegic is the combination of external rotation in the legs with the very difficult valgus foot (Figure 7); difficult usually because of the combination of a progressively valgus hindfoot with a supinated and abducted forefoot, and relative weakness of the invertor muscles. This is sometimes called a valgus-ex-equino deformity in the belief that the valgus arises secondarily to the tight tendo-achilles. In practice, however, lengthening of the tendo-achilles rarely improves the condition and can make it worse. The deformity tends to progress until it is very severe. It can be almost impossible to correct even by bony procedures such as triple arthrodesis and preventive surgery performed early is likely to give the best results. Unfortunately at present there is no general agreement on the best method of dealing with this problem. A wide variety of procedures have been used, one of the most popular being the Grice sub-

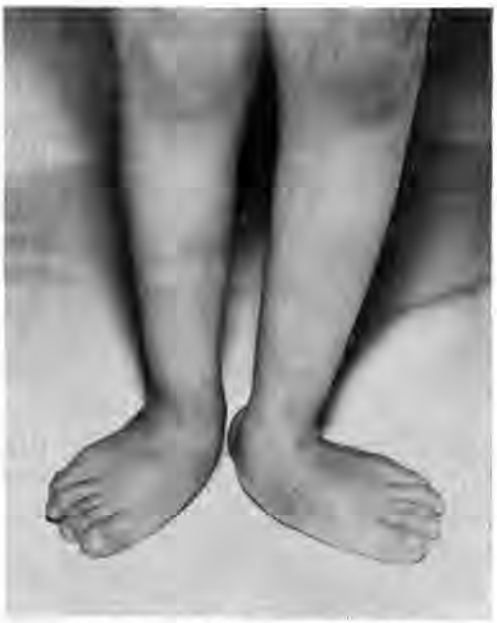

Fig. 7. Valgus foot in spastic diplegia.

talar arthrodesis (Grice, 1952), a procedure which, in cerebral palsy, frequently fails to prevent progression of deformity. In my experience, one of the most successful procedures, provided it is performed early, is to transfer one of the peroneal tendons to the medial side of the foot, usually into the tibialis posterior, and combine it with lengthening of the other peroneus tendon. Also useful in the carefully selected case is the medially based, closing wedge os calcis osteotomy which realigns the tendo-achilles so that it tends to pull the foot round into varus. However, both these procedures carry the disadvantage that they are somewhat unpredictable, and the end result may be a severe deformity in the opposite direction. Whatever method of correction is used, it is rarely possible to avoid the use of below-knee braces to control the tendency of the foot to sag into valgus. This is one of the few situations where bracing is necessary.

There has for a long time been a tendency to regard a triple fusion as being the ultimate answer to any foot deformity, but experience of this procedure suggests that it can be extremely difficult to perform in the badly deformed foot and an inadequate correction may result. It is therefore important to strive to obtain a well shaped plantigrade foot as early as possible and to maintain this by repeated surgery if necessary.

In summary then these constitute some thoughts on the principles of orthopaedic surgery in the treatment of cerebral palsy illustrated by their application to some of the more common problems. Nevertheless the variety of handicaps and deformities in these children is enormous and it must be appreciated that even when considering the lower limbs only, as I have tended to do in illustrating these principles, the range and scope of surgery is considerable. The results of treatment in these children are extremely gratifying and we must hope that, in well-run cerebral palsy clinics, the days have now gone when we see grotesquely deformed children with pressure sores and pain, and no hope of achieving their true potential level of function.

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