THE PROSTHETIC MANAGEMENT OF CONGENITAL DEFORMITIES OF THE EXTREMITIES

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Congenital deformities of the extremities may come within the limb fitter’s sphere after amputation or when, without amputation, there is sufficient shortening or deficiency to allow the fitting of an artificial limb to the deformed limb. The classification of these cases is difficult because they present wide variations in anatomy, but certain broad groups emerge in relation to prosthetic management. The material for this paper was gathered from a series of 166 cases with a follow-up of up to eight years, together with many others treated by the Limb Service both at Roehampton and in the provinces. The numbers of each type of case are shown in parenthesis in each section, but they are not to be taken as indicating the incidence of these conditions, because the series is composed of highly selected cases.

CLASSIFICATION

LOWER LIMB DEFORMITIES

Deformities primarily affecting the foot (two cases)—These deformities are not common. In appearance they may resemble a surgical partial foot amputation. Rudimentary toe buds may be present. There may be some shortening of the limb as a whole, and a degree of equino-varus has been seen in association with the foot deformity, as after surgical amputations at this level. One or more digital rays may be absent (Fig. 1).

Deformities primarily affecting the leg (twenty-seven cases)—The common deformity in this region is absence of the fibula. There is usually marked shortening, some of which may be due to associated shortening of the femur. The hip and knee are usually sound, but they may show flexion deformity, and genu valgum is not uncommon. Half the pelvis, and the long bones on the affected side, are sometimes underdeveloped. The ankle is usually unstable, with varying degrees of equino-varus. The tibia often shows deformity which may be anything from a slight bowing, usually forwards, to complete pseudarthrosis. The foot may be almost normal or it may be deformed. Commonly one or more digital rays are absent, there may be syndactylism of two or more toes, and the tarsus may be deformed and incomplete. A skin dimple can usually be found at the front of the leg, often at the junction of the middle and lowest thirds, which is the common site of bowing of the tibia.

Occasionally it is the tibia that is absent. Although the fibula may hypertrophy considerably, the limb is unsatisfactory. The ankle is grossly deformed and unstable, and perhaps the knee likewise, and weight bearing cannot be tolerated (Figs. 2 and 3). It is usually advisable to amputate the limb in these cases. The skin dimple is usually present.

FIG. 1
Congenital deformity of foot resembling Chopart’s amputation. Note equino-varus and toe bud. (Mr Leon Gillis’s case.)
Deformities primarily affecting the thigh (nineteen cases)—These deformities are also relatively common. The femur is shortened and there is often a severe flexion deformity of the hip and knee, so that the thigh is held much in the position seen in quadrupeds. The shaft of the femur may be deficient in various ways (Fig. 4). It may be represented by a relatively normal lower end, tapering away to a point a few inches proximally. The head and neck may be absent or underdeveloped, and there may be severe coxa vara. There is sometimes a pseudarthrosis of the femur. The hip may have good function, or it may be rudimentary and represented by a shallow or absent acetabulum with the attenuated upper end of the femur lying more or less in its proximity. The skin dimple can usually be found.

The knee is often sound, but flexion deformity is common. The leg and foot are usually normal. These limbs are usually surprisingly strong, and, even in girls, may be strikingly hirsute in comparison with the other limbs.

Congenital amputation stumps—A congenital deformity having the appearance of a surgical amputation stump is rare in the lower limb, but common in the upper. When it occurs in the lower limb it usually takes the form of a below-knee stump which may be very short. The epiphysis may be present and the anatomy of the hip, thigh and knee is usually fairly normal, but congenital dislocation of the hip in association with a congenital through-knee "amputation" has been seen. Toe buds are common. This type of deformity, in the lower limb, has not been seen as a solitary defect (Fig. 5).
Multiple and bizarre deformities (thirty cases)—The deformities described above may exist singly or in combination. They may be confined to one limb. There may be a concomitant deformity of the upper limb, in which case it is usually, but not necessarily, the same side that is affected. Both legs may be deformed, in which case the deformities are seldom symmetrical (Fig. 6). One patient has been seen with total absence of both lower limbs, the asymmetry in this case being represented by slightly greater development of one side of the pelvis and greater bulk of the overlying soft tissues. Two children have "amputation-stump" deformities of all four limbs.

"Constriction band" deformities (four cases)—Some limbs show appearances which suggest that they were caused by constriction. A constriction ring is present, the limb distal to the ring being atrophic to a varying extent (Fig. 7). The site of the constriction ring is not constant. There may be loss of the extremities of the limb; thus the toes may be absent or represented by buds, or there may be greater deficiencies up to an amputation-stump deformity. The circulation of the distal part of the limb may be precarious. It seems possible that the mechanism producing constriction band deformities also produces the amputation-stump deformity.
Deformities of the lower limbs, with tubing applied to the sulci to demonstrate the possibility of the umbilical cord's having produced the constrictions. (Mr Leon Gillis's reconstruction.)

deformity if the constriction is sufficiently severe (Fig. 8). Absence or deficiency of the long bones has not been seen in association with constriction band deformities in this series.

Amputation necessitated by spina bifida (seven cases)—Although these are not cases of deformities of the extremities, they arise out of a congenital deformity and are conveniently included in this paper. When amputation is resorted to in these cases it is commonly on account of trophic ulceration of the feet. These patients present one of the most difficult problems to the limb fitter. The stumps often lack sensibility completely or in part, and the incontinence that is so often present adds to the difficulties of providing a prosthesis that will not cause chafing and further ulceration of the stumps. When the skin overlying the ischial tuberosities is also anaesthetic the problem may be beyond solution. Because of these difficulties in limb fitting, amputation should be undertaken only as a last resort, and the prognosis in respect of function with artificial limbs should be guarded (Figs. 9 and 10).

Severe constriction band deformities of both lower limbs. Relief of constriction by plastic surgery was required because of circulatory deficiency before a prosthesis could be tolerated on the left side. (Mr R. J. V. Battle's case.)
Figure 9—Congenital absence of sacrum and lower lumbar vertebrae. There were trophic lesions of lower limbs. Treated by amputation of the right leg below the knee and disarticulation at the left knee. Patient is now walking on more or less conventional prostheses. (Mr Vincent Snell's case.)

Figure 10—Same patient, showing preliminary pylons. Note extreme development of shoulder girdle and arms. This boy ultimately mastered articulated legs.

Figure 11—Through-elbow amputation type deformity. Note hand bud. (Mr E. E. Harris's case.)

Figure 12—Severe bilateral deformities of arms. Fusion of elbow joints and shortening. Note absence of opposition of thumb and fingers. Skin dimple is seen on the right arm. (Sir Arthur Porritt's case.)
UPPER LIMB DEFORMITIES

Congenital “amputation stumps” (twenty-four cases)—Congenital absence of the distal part of an upper limb is not uncommon. Since the deformity resembles that resulting from surgical amputation, most of the patients are sent to a limb-fitting centre. This may account for the preponderance of this type of deformity among our cases. Thus out of twenty-eight new patients with congenital deformities referred to Roehampton in the past six months, fifteen had this type of arm deformity. In order of frequency the common sites of congenital amputation are: below elbow, through wrist, carpal or metacarpal, through elbow, above elbow, shoulder (Fig. 11). Most of these cases are treated by routine methods and are therefore not included in the present series.

Deformities affecting the hand (fifteen cases)—These usually take the form of syndactyly, microdactyly, etc., and seldom come before the limb fitter.

Multiple and bizarre deformities (sixteen cases)—The common feature of most other arm deformities is fusion of the elbow joint with varying degrees of shortening. The hand may be almost normal or may show deficiencies in digital rays, especially of the thumb. There may be only one digit, or there may be an aberrant digit sticking out at a grotesque angle. Syndactyly is common. The skin dimple can often be detected. These deformities are incapacitating, especially as they are often bilateral. Fortunately they are uncommon (Fig. 12). Complete absence of both upper limbs has been seen.

GENERAL MANAGEMENT

Children exhibiting these deformities are nearly always of high intelligence and often they are strikingly handsome. The sexes seem to be equally affected, and all social grades are equally at risk. Little evidence in favour of a hereditary trend has been found. If confirmed, this observation puts these deformities in sharp contrast to such conditions as syndactyly and polydactyly, in which a hereditary influence is accepted (Barsky 1951, Bagg 1929) and a further study of this important aspect of the problem is being made.

Psychological adjustment to these deformities is usually good, especially when a reasonably good aesthetic and functional result is obtained by limb fitting. A few children have shown aggressive or exhibitionist traits, and one child developed a facial tic. Such reactions usually resolve when the child goes to school. Unless deformities are grotesque or severely disabling the children should go to a normal school, where their high intelligence stands them in good stead.

Abnormal psychological reactions in the parents of deformed children are more common. There have been instances of suicide of one or both parents after the birth of a severely deformed child. Marriages have broken down through one parent’s implying that the deformities in the child were the result of bad stock on the other parent’s side or through allegation of cruelty during the pregnancy, or from morbid obsessions of guilt. Some parents have deserted a deformed child or simply refused to have anything to do with it, necessitating its being cared for in a nursery or a school for the disabled. In most cases, however, the parents accept the child and their responsibilities to it and manage it well with the right mixture of sympathy and firmness. They are encouraged to make little of the deformity and to treat the child as normal.

PROSTHETIC MANAGEMENT

LOWER LIMB

Every patient with a congenital deformity presents an individual problem. Detailed clinical and radiological examination is necessary to assess the capabilities of the deformed limb, the range of movement of the joints and the capacity for weight bearing. In general, if there is enough shortening to allow of the fitting of at least an artificial foot, most of these patients can be fitted with an extension prosthesis without surgical intervention, and in many
cases this will give the best functional result. In some cases operation to correct flexion or other deformity may have to be considered before limb fitting is undertaken. In others an amputation may be indicated.

Limb fitting for the lower limb is begun as early as possible, ideally when the child begins to make efforts to stand. At this stage, when the limbs have not been subjected to weight bearing, the deformities are probably not in their final state. It may seem politic to amputate, for example, a foot in equino-valgus with absence of the fibula. But an extension prosthesis can usually be fitted, involving nothing that is irrevocable, and amputation can be resorted to later if the functional or aesthetic result is unsatisfactory. It is surprising how many parents think that the deformed leg will grow to be normal in time if it is left to itself, and no amount of explanation will disillusion them. If the leg is amputated in infancy they may never accept that it was done for the best, and may continue to believe that all would have been well had the limb been left alone. A similar reaction has been seen in the children themselves. One child seen at the age of sixteen years had had below-knee amputations of both legs in infancy for deformities. Prostheses had been fitted but she had not done well with them. She was sullen and unco-operative, and it transpired that she had conceived a hatred for her mother for bringing her into the world with deformities, and for her father for agreeing to the amputations, for she thought that the limbs would have come right in time.

When contemplating amputation, therefore, the surgeon should satisfy himself that the operation will help the patient functionally or psychologically and should resist the natural temptation to remove freak anatomical structures and leave a stump of serviceable appearance. In young children amputation should be resorted to only when there are clear indications that the child will gain in function. Very rarely psychological reactions may make operation necessary, but these can usually be overcome by suitably reassuring the child and, more particularly, the parents.

Nevertheless, a conservative attitude can be maintained too long, as the following case illustrates. A girl had been in our care wearing an extension limb for congenital absence of the fibula since the age of eight. The functional result was excellent, but aesthetically it left much to be desired. Through our failure to recognise that she was a precocious child she became self-conscious of her appearance, refusing to wear anything but slacks, and became unruly and unmanageable. A Syme’s amputation was ultimately agreed to at the age of fourteen; the aesthetic result was good, and psychologically the child was restored to normal. This child would have had a happier childhood if her parents’ request for amputation had been met earlier.

It should not be forgotten that a girl reaching maturity is at a serious disadvantage in her prospects of marriage if she has a freakish deformity and, although an amputation is also a handicap in this respect, a relatively normal looking stump may be more acceptable to the prospective husband than a deformity which is more or less bizarre. It is probable that this is in the minds of some of the girls who seek amputation for cosmetic reasons as they grow up.

It follows that each case must be treated on its merits and, in borderline cases, the greatest care must be taken in balancing the functional and psychological requirements. Nevertheless, especially in the younger patients, it is felt that nothing can be lost by an attempt to fit an extension prosthesis in the first instance, resort being made to amputation only if the results fall short of requirements. Whenever possible surgery should be deferred until the child is old enough to share in the decision.

The extension prosthesis is constructed as follows. A boot, into which the natural foot is fitted, is made to a cast of the foot in a position of equinus and fixed on a platform. The greater the equinus the less obtrusive is the prosthesis, but care should be taken not to force the position to the extent that the heel slides off the platform. Some assistance can be obtained by fitting the toes in slight dorsiflexion. Below the platform, as much as is possible of a conventional prosthesis is applied by means of side steels in much the same way as in the
O'Connor appliance. The side steels may be extended upwards to carry a thigh corset which can afford ischial bearing if required. Knee joints can be fitted to the side steels and such devices as T-straps and patella straps applied. The whole appliance is made and set up to a cast of the deformed leg. For young children the lower part of the appliance is made from wood, with a fixed ankle and a felt forefoot. For adolescents and adults a metal base may be used, and in a few cases it may be possible to enclose the deformed foot more or less completely within the shell of a metal shin with excellent aesthetic effect. One such patient was enabled to achieve her ambition of singing on the concert platform, which she had been too sensitive to do before.

**Deformities primarily affecting the foot**—These limbs can usually be fitted with one of the conventional prostheses for Chopart's amputation, with relatively minor modifications. When there is shortening of the leg as a whole it may be necessary to make a modified platform limb. This gives good function but is unsightly because of the bulk at the ankle.

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**FIG. 13**
Congenital absence of fibula. Drawing prepared by superimposing tracing of radiograph on sketch of prosthesis. The boy came third in his school high jump in open competition with normal children.

**FIG. 14**
Old-standing tuberculous hip with premature epiphysial arrest and backward subluxation of the knee. The hip was ankylosed. Fitted with corset-top caliper with free knee joints mounted on platform prosthesis. Cross knee strap and calf band tended to correct knee deformity.

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**Deformities primarily affecting the leg**—These deformities are usually associated with sufficient shortening to allow of the fitting of an extension prosthesis. The capacity of the limb for weight bearing must be assessed; if it is in doubt an ischial bearing corset should be fitted, although it is usually possible to retain knee function by fitting joints to the steels (Fig. 13). The varus deformity that is commonly present at the ankle can be controlled by a T-strap. Tibial bowing, flexion contractures at the knee and hip, and genu valgum frequently improve under the stimulus of weight bearing, and may disappear entirely. When the knee and hip are sound the boot does not need to be extended beyond the tibial condyles, which can be used, by means of a blocked leather boot, to accept some of the weight load if this is desirable. Genu valgum can often be helped in young children by a wedge inserted between the ankle and the foot of the prosthesis.

The functional result with this type of deformity is uniformly good after satisfactory fitting with a prosthesis. Many patients can even run without a limp. One boy was placed third in his school high jump in competition with normal boys. The aesthetic result is less
satisfactory, and amputation at the ankle may be required in girls at a later stage for this reason alone. Patients with a tibial pseudarthrosis which fails to unite probably require amputation through the pseudarthrosis to get satisfactory function.

Patients with shortening from arrest of growth after poliomyelitis, tuberculous disease or other causes can be fitted with a similar extension prosthesis. When control of the hip is weak it is possible to fit a rigid hip joint and pelvic band to the top of the outer side steel. There are fifteen such cases in this series (Fig. 14).

Some cases of leg deformity have been referred to the limb centre after amputation. This usually takes the form of a modified Syme's amputation, the foot being removed at the ankle joint. The resulting stump is usually satisfactory and can tolerate end bearing. The end of the stump is not so bulbous as is a true Syme's stump. Patients with this type of amputation are usually fitted with the metal enclosed type of Syme's appliance, a fixed ankle and felt foot being used while the child is small. The functional results are comparable with those of an extension prosthesis, but if the fibula is absent there may be difficulty in preventing the prosthesis from rotating on the stump. It must be admitted that the aesthetic result is better and, indeed, may be perfect. Occasional difficulty has been experienced with young children who have been subjected to amputation in that the memory of painful experience remains and they are frightened and unco-operative. This can usually be overcome by the removal, by those in attendance, of the white coat and by suitable distractions in the way of playthings and sweets. If the child will not permit a cast to be taken at the first visit a second attempt a few weeks later is usually successful.

When the tibia is absent it is seldom possible to conserve a functional knee joint, and a disarticulation at the knee will probably be required, followed by the fitting of a conventional prosthesis.

**Deformities primarily affecting the thigh**—These cases provide a more difficult prosthetic problem. The flexion deformities of hip and knee are such that the knee is usually more or less at the same level as the hip and lying anterior to it, so that the axis of the leg is anterior to that of the bodily centre of gravity. If a platform prosthesis is fitted in the axis of the leg it is unstable and tiring to manage, and a free swinging artificial knee cannot be controlled.
If the whole weight is carried by the foot on the platform there is a strong tendency to "jack-knifing" at the hip and knee. The movement of flexing and extending the shank on the trunk is a complex one commonly taking place partly at the hip but mostly at the knee, and at the pseudarthrosis if present. If it is necessary to use an artificial hip joint its best position can be determined only by trial and error.

It follows that the extension prosthesis for deformities of this type must usually afford ischial bearing. The side steels should be allowed to incline slightly backwards from above downwards so that, at the level of the platform, they are in the plane of the axis of the centre of gravity. The natural heel is then usually at about the same level as the normal knee. Knee joints can be fitted to the steels at this level, the boot on its platform being attached to the upper section of the steels (Fig. 15). Function with this type of prosthesis is usually quite good, and is comparable with that obtainable by a patient with disarticulation at the knee. Aesthetically the anterior position of the knee produces a bulge which may be more or less conspicuous according to the length of the femur. In a sitting position the toes tend to project forwards and show through the trouser leg or below the skirt. This can be minimised by setting the artificial knee joints as low as possible (Fig. 16).

In some cases the prosthetic problem has been simplified by surgical correction of the deformities of the hip and knee flexion, with arthrodasis of the knee in extension. This may give an excellent limb in good alignment, with robust function on a platform prosthesis.

Sometimes a Syme's amputation, either alone or in association with a corrective operation, has been used. The stump can then be fitted into a conventional thigh prosthesis if the socket is modified appropriately, but care must be taken that the socket is aligned in the container in such a way that the axis of the prosthesis is in the plane of the axis of the centre of gravity rather than in the line of the tibia. The aesthetic result of this procedure is good, but it is doubtful if the functional result is ever so robust as that obtained with the extension prosthesis.

Van Nes (1950) described another method of treating these cases. Arthrodasis of the
knee is performed and a femoral osteotomy is done. The distal fragment is rotated through 180 degrees and retained in this position while the osteotomy unites. This brings the heel round to the front. The foot is fitted into a specially fitted socket within an otherwise conventional below-knee limb prosthesis. The patient’s ankle then acts as a knee to control the movements of the artificial shin. Aesthetically and functionally the possibilities of this technique seem good if the full 180 degrees of rotation can be ensured, and if there is no residual flexion deformity of the hip. I have seen only one patient so treated. In this case the rotation was a few degrees short of 180 degrees, and after a subsequent accident in which the femur was refractured union occurred with still more loss of rotation, resulting in difficulties in the alignment and "tracking" of the prosthesis. There was a slight residual flexion deformity of the hip, and the boy never walked with great confidence and could not part with his stick. It is emphasised that the relative failure of this case was due to alignment

![Image](image-url)

**Fig. 19**

The first set of prostheses supplied at the age of two years to the patient shown in Figure 18. The type of walking stick with flat base illustrated has proved of value for young children.

problems and does not necessarily imply that the principle is unsound. It was interesting in this case to note that the calf muscles took over the function of the quadriceps in the rhythm of gait immediately and without extensive re-education. Presumably the reflex originates in the plantar sensory nerve endings (Whillls 1953).

**Congenital amputation stumps**—These can be fitted with conventional limbs and present no special difficulties unless there are complicating factors such as congenital dislocation at the hip. The latter will probably necessitate the provision of an ischial bearing limb and, if a prosthetic hip joint should be required, the correct position for it may be difficult to determine and must be decided by trial and error.

**Multiple and bizarre deformities**—These produce problems of great complexity and each case must be taken on its merits. When both lower limbs are short it is often possible to begin prosthetic work on the shorter of the two. The longer limb being fitted without extension, if necessary with a surgical boot, and possibly with a caliper if required for stability. When
mastery of this apparatus is achieved, progression is made by fitting an appropriate extension prosthesis to the longer limb to bring the limb into proportion with the general bodily physique, the prostheses for the shorter limb being lengthened correspondingly (Figs. 17 to 19).

For example, the patients with one limb represented by a foot located close to the buttock, and the other limb by a deformity of the thigh, have undergone the following programme. First the foot on the short side was fitted on a specially made ischial bearing socket within an otherwise conventional thigh pylon. In one case in which the small foot had, unfortunately, been amputated, it was necessary to use a tilting table pylon. The length of the pylon was adjusted to permit of weight bearing on the other limb with no prosthesis equipment apart from a surgical boot and ischial bearing caliper if required. When stability and control had been achieved the pylon was replaced by a fully articulated conventional prosthesis, and an extension prosthesis was fitted to the other limb deformity. The raising to full stature was done in several stages. The results in these cases have been very gratifying. Some patients are even able to go to normal schools.

Constriction band deformities—These cases are treated on similar general principles. Thus, a patient with ring constrictions at both ankles was fitted with a simple extension prosthesis on the shorter side and was beginning to walk, but ulceration from circulatory deterioration made it necessary to have a " Z-plasty " done to improve the blood supply. It is probable that this patient will require amputation eventually.

Spina bifida with amputation—Amputation stumps in association with spina bifida are almost always partly anaesthetic, and it is necessary to relieve these areas of weight bearing and friction. The general principles of limb fitting are applicable. The below-knee stump must be protected by full ischial bearing when sensibility in that area is adequate, and a loosely fitting felt-lined slip socket suspended to the stump by elastics protects the anaesthetic stump. When there is incontinence the prosthesis must be protected against the corrosive effects of urine. The leather-work is treated by coating with leather lacquer. Various plastic coatings have been tried, but it is difficult to obtain satisfactory bonding and the plastic skin has tended to peel or flake. Recent work suggests that silicones may be of value for this purpose. Polythene thigh corsets have been used with some success. The steels can usually be protected adequately by heavy plating. Duralumin is very susceptible to corrosion. A good cellulose paint will protect the metal provided it remains unscratched. Anodizing is also effective so long as the skin so formed remains intact, but this method is expensive and scratches will not readily be seen until corrosion has set in. Stainless steel is also a possibility but it is difficult to work.

UPPER LIMBS

It is not our practice to begin limb fitting at such an early age in upper limb cases as in lower. The instinct to walk is practically irrepressible, and use can be made of this in fitting a prosthesis to the lower limb at about the age when the child could normally be expected to start walking. In the case of the upper extremity, however, the deformed arm or stump is used freely and is usually sufficiently dexterous to meet the simple needs of the very young child, who is not self-conscious of the deformity. Nevertheless we regard it as important to have the child fitted with the prosthesis and accustomed to its use by the time he goes to school. At this stage he is plunged into a new environment, and to reduce his inevitable sensitivity to comment by his fellows, he should look as normal as is possible. Apart from this we have no hard and fast rules but, by keeping these patients under observation, begin prosthetic work when the child's intelligence has developed sufficiently to co-operate well. The youngest child that we have fitted with a functional arm prosthesis was two and a half. We have known instances in which premature limb fitting has caused the child to form a revulsion from the prosthesis and to persist in refusing to wear it through the years.

The usual procedure is to fit first a simple arm of largely aesthetic value, and to progress to a more functional arm as intelligence develops.
Hand represented by single powerful digit. Palm case with artificial thumb was provided to give opposition, but the appliance was of little practical value.

**Congenital amputation stumps**—These can be fitted with the conventional prosthesis appropriate to the length of stump, and no special difficulties arise.

**Deformities affecting the hand**—These seldom come within the province of the limb fitter unless the hand lacks the power of grasp. In these cases it may be possible to fit a palm case carrying an opposition plate like that used for a partly mutilated hand. In this way pincer function can be provided (Fig. 20). These cases are uncommon, and they often give disappointing results. The patient usually has become so expert in using the digits he possesses that he finds any appliance an encumbrance. Moreover some patients, after attempts at surgical reconstruction, have scars in awkward situations, possibly hyperaesthetic and sometimes associated with contractures. In consequence of such unsuccessful surgery the patient’s morale may be low.

Some patients in this group come for the provision of a purely aesthetic prosthesis, for instance to replace absent digits. These cases, too, may give disappointing results because the structure of the natural hand is often such that the prosthesis cannot be mounted to look natural and inconspicuous: in any case,
the prosthesis is inevitably an encumbrance, and it is often quickly discarded (Fig. 21).

**Multiple and bizarre deformities**—This group provides the most difficult problems in upper limb prosthetics. The deformity commonly affects both sides. The general appearance is usually more or less grotesque and the patient is inevitably sensitive about this. The deformities are usually such that it is impossible to make a prosthesis hang in a natural position. The patient has usually achieved such skill with the deformed hands that the application of prostheses, which necessarily enclose the hands, so impairs function at first that he feels frustrated and may abandon the attempt. From this point of view, when both arms are deformed, it is advisable to approach the prosthetic management on one side at a time, and the less useful side should be attempted first.

The fused elbow joint presents problems of its own. If the length of the arm as a whole is full or nearly full the patient is prevented from reaching his mouth, and indeed from reaching the head and neck and much of the upper half of his body. A prosthesis cannot provide this function without being disproportionate. There is no satisfactory prosthesis for a deformity of this sort, and the provision of a surgical pseudarthrosis must be considered, especially when the deformity is bilateral.

Three unusual cases of severe double deformities have been seen. In each the long bones of one arm were shorter than in the other, but the anatomical features were substantially the same. The hands lacked one or two digital rays but otherwise were fairly normal. They articulated through the wrist with a short forearm composed of a single bone which probably represented the upper end of the ulna fused to the lower end of the radius. An elbow joint
of sorts was present, and a short humerus terminated abruptly at its proximal end, where it lay some distance below an indefinite glenoid. There were sharp contractures of all the joints, so that the arms were held in much the position of a dog's forepaws when begging. These patients were highly intelligent and used their short limbs remarkably well. They have been fitted with prostheses of definite, if limited, usefulness (Figs. 22 and 23).

Three patients have been seen with total or almost total absence of the upper limb. They had acquired astonishing skill in the use of their feet: one woman did exquisite needlework in this way. She did not persevere with the prostheses that were supplied because their function was limited and she was, at the age of forty-two, too set in her ways to learn. The second patient is fully rehabilitated and in employment as an accountant. The third is aged fourteen. Fitting is being undertaken and is promising well. Pectoral cineplasty has been considered in this case but, in addition to parental resistance to the consequent mutilation, it was not felt to offer much success because the chest was narrow and the range of movement of the tunnel would have been small.

Flail arms (eight cases)—Prosthetic technique can be applied with benefit in certain cases of flail or partly flail arms. For instance, it may be possible to provide voluntary flexion of a flail elbow by using a harness like that used for the above-elbow prosthesis mounted to an elbow cage. If the hand is functionless, a device can be fitted to a suitable palmar splint so that a split hook or other prosthetic tools can be carried and operated. Control of the scapular muscles is, however, a prerequisite of success in these cases.

It will be realised that the types of deformity described above are by no means comprehensive and that the possibilities of individual variation are infinite. Each case must be taken on its merits, and many demand the utmost technical ingenuity. The closest liaison between surgeon and prosthetist is demanded at all stages in the construction of the prosthesis and, when difficulties arise, an orthopaedic opinion should be sought on the possibility of improving the limb surgically. The plastic surgeons are able to help in selected cases.

SUMMARY

1. Congenital defects of the extremities are described. Although the detailed anatomy is infinitely variable, a broad classification in relation to prosthetic management has been suggested.
2. Most patients with these deformities can be fitted with a prosthesis without major surgical intervention. With this they will have at least as good function as they would have after amputation. A plea is made for a conservative attitude in this respect. It is suggested that recourse to amputation should be confined to cases in which prosthetic equipment falls short of functional and cosmetic requirements, and that, when possible, it should be deferred until the child is old enough to share in the decision.
3. The prostheses applicable to the various types of deformity are briefly described.
4. The application of similar techniques to cases of acquired shortening is mentioned.
5. The incorporation of certain features of artificial arms in flail arm splints is discussed.

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