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THALIDOMIDE BABIES: MANAGEMENT OF LIMB DEFECTS

BY

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From observations of the recent cases of congenital limb deformities attributed to thalidomide administered during the early weeks of pregnancy (that is, fifth and sixth weeks) it would seem that there is a fairly clearly defined clinical syndrome (see Smithells, 1962; Leck and Millar, 1962).

There is, commonly, a haemangioma situated centrally on the forehead. Often another is seen on the upper lip and sometimes on the cheek or the tip of the nose. These fade completely in time. Associated with this phenomenon is a depressed bridge of the nose, referred to as "saddle nose." Rarely there is microtia, and this may be associated with a partial facial palsy. Coloboma affecting one or both eyes is also sometimes seen. All the limbs are usually affected and phocomelia is the commonest type of deformity, there being various degrees from complete absence of the long bones, with one or two fingers at shoulder level, to limbs of normal length with hands having four fingers and absent thumbs or rudimentary thumbs (Figs. 1–3). This type usually has a defective or absent radius. In the upper limbs the defect always seems to be "radial"—that is, if only one finger is present it is the little one, then little and ring finger, etc. Similarly in the lower limbs there is often the long-bone defect and defects of toes, although polydactyly is not uncommon. The deformities are usually symmetrical regarding length, but often one side is slightly more developed than the other—that is, there may be three fingers on one side and two or four on the other. Upper limbs may be very short, while lower limbs are either normal or only very slightly affected. This is far more common than the reverse. More rarely, an infant is seen with bilateral amelia (absence of limb or limbs) and the other limbs normal, or with bilateral amelia and bilateral phocomelia. A single limb defect is quite rare.

There seems to be no mental retardation in the majority, although it has been reported. Defects of the cardiovascular system, urogenital system, and intestinal tract are moderately often seen.

Principles of Management.—The orthopaedic management of the limb deformities of thalidomide babies is the same as that for similar deformities due to other causes. Their treatment follows well-established principles (Gillis, 1956). It consists essentially in (1) correction of deformities and contractures by conservative means, (2) surgery to salvage badly deformed limbs, or amputation, and (3) the uses of prostheses.

Conservative Correction of Deformities

Of prime importance is the orthopaedic correction of deformities and contractures. Manipulative measures should be undertaken at a very early age in order to produce anatomical alignment and cosmetic re-position as far as possible. Serial manipulative plasters followed by plastic retentive splintage are suitable for mobile deformities. In no circumstances should amputation be done before considering what can be accomplished by ordinary orthopaedic care, by adequate splintage, and by prostheses.

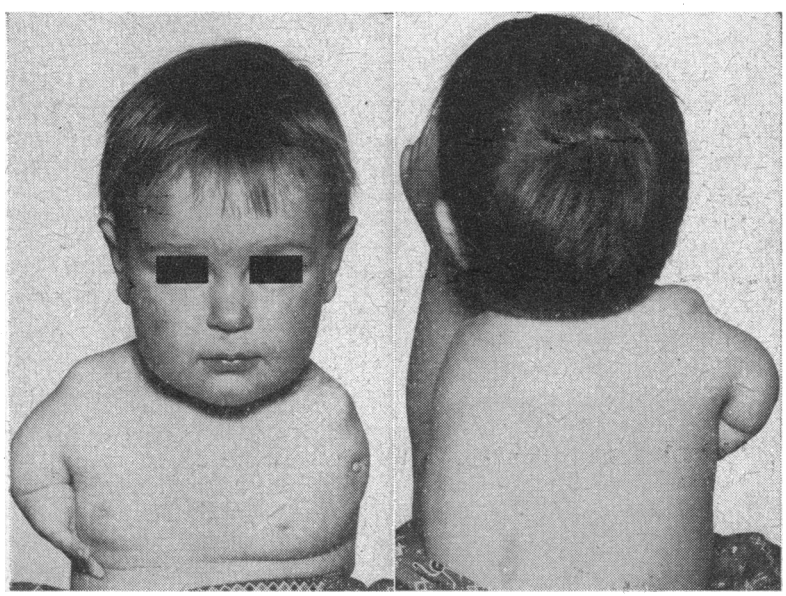

FIG. 1.—Boy aged 18 months, whose mother gave a history of having taken thalidomide during early pregnancy. Note "saddle nose," and haemangioma situated on right cheek. There is a complete absence of left upper limb; on right side he has a rudimentary humerus, absent radius, and two rudimentary digits.

The object of splintage is to correct deformities in the developing bones and joints. The object of an appliance, such as a patten, is to initiate walking and desirable movements before a semi-final decision is made regarding the desirability of supplying an artificial limb. The object of an artificial limb in these cases is (1) to conceal the deformity, (2) to extend a short limb and enable natural walking to take place, or (3) a combination of both. Such a limb is called an extension prosthesis or platform limb. It is a type of appliance which is designed to conceal deformed short limbs due to congenital abnormality, poliomyelitis, etc. The special features of the limb will vary according to the particular deformity concerned, but the general principle is as follows: a boot is constructed into which the natural foot is fitted (Fig. 4). This boot holds the foot in a position of equinus and is fixed on a platform at the required distance from the ground. The greater the degree of equinus the less obtrusive is the

prosthesis. Below the platform as much as is possible of a conventional prosthesis is attached by means of side steels. The side steels may be extended upwards to carry a thigh corset which can afford ischial-bearing if required.

Knee-joints and such devices as T-straps to correct deformities, or knee-straps to maintain the correction, are usually fitted to the side steels. For young children the lower part of the appliance is usually made of wood with a fixed ankle and a felt forefoot. For adolescents it may be possible to enclose the deformed limb and foot in a plastic or metal shin, which looks natural.

Role of Surgery

Once manipulative measures for correction have produced anatomical re-alignment so far as is possible, surgery can be planned. This will depend on the type and state of the residual deformity.

To make limb-fitting more successful, or sometimes even only practicable, badly deformed and grotesque limbs can be improved by reconstructive surgery in order to get better alignment, to correct deformities, and to increase length. It should be stressed, however, that because of the variety of the deformities there can be no fixed procedure beyond arthrodesis of joints or osteotomies to restore alignment.

Where a congenital dislocation of the hip is present in an abnormal limb, this will be treated according to the accepted

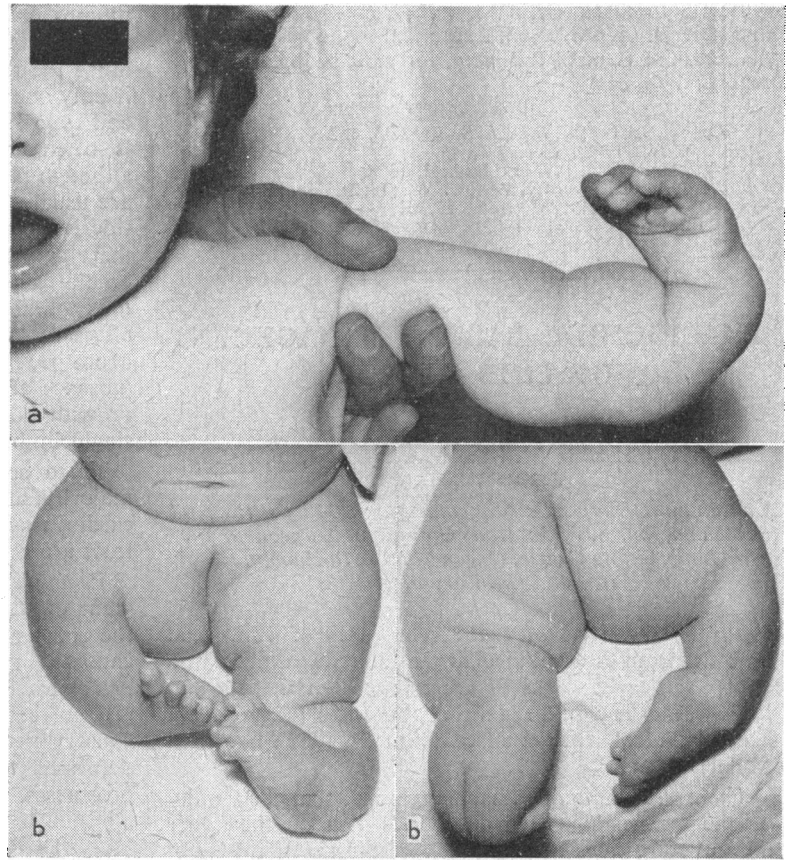

FIG. 3.—Girl, aged 1 year, whose mother states she had taken thalidomide for 12 months (before and during pregnancy). (*a*) The left arm shows short deformed radius and ulna fused at the upper end, and only four metacarpals. (*b*) Shows right leg with deformed femur. The upper end is not clearly seen, but fragmented epiphysis is present. Fibula is missing. Left leg has head of femur epiphysis missing. The tibia is short and deformed, the fibula is missing, and both hips, knees, and ankles are dislocated.

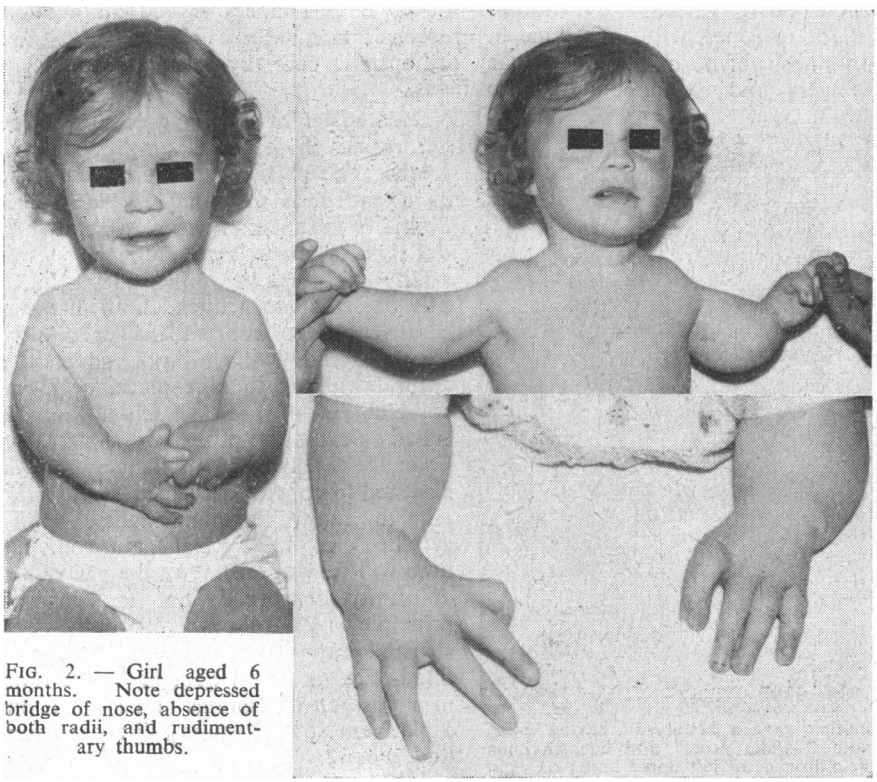

FIG. 2. — Girl aged 6 months. Note depressed bridge of nose, absence of both radii, and rudimentary thumbs.

methods of reduction, or operative intervention if necessary. Operative procedures like tenotomy, excision of joints, osteotomy, or osteoclasis are undertaken where necessary.

Children readily adapt themselves to wearing artificial limbs and splints, so that it is sometimes possible to accelerate their progress by operative treatment, even amputation. Surprisingly good functional and cosmetic results can be achieved by early and well-considered splinting or limb-fitting.

It is accepted that the case presented for amputation and limb-fitting is one where the surgeon has satisfied himself that reconstructive operations, such as limb-lengthening and bone-grafting, will not prove satisfactory or, if they have already been performed, have failed to produce a satisfactory limb. In these cases where amputation is of necessity

performed at an early age, overgrowth of the cut end of the bone in the stump may produce problems as the child grows. Revision of the stump often becomes necessary, even though the bone has been covered with soft tissue and skin. The younger the child at amputation the more liable he is to develop a "conical stump." However, this problem does not arise where there is congenital absence of a part of a limb which has had no surgical intervention.

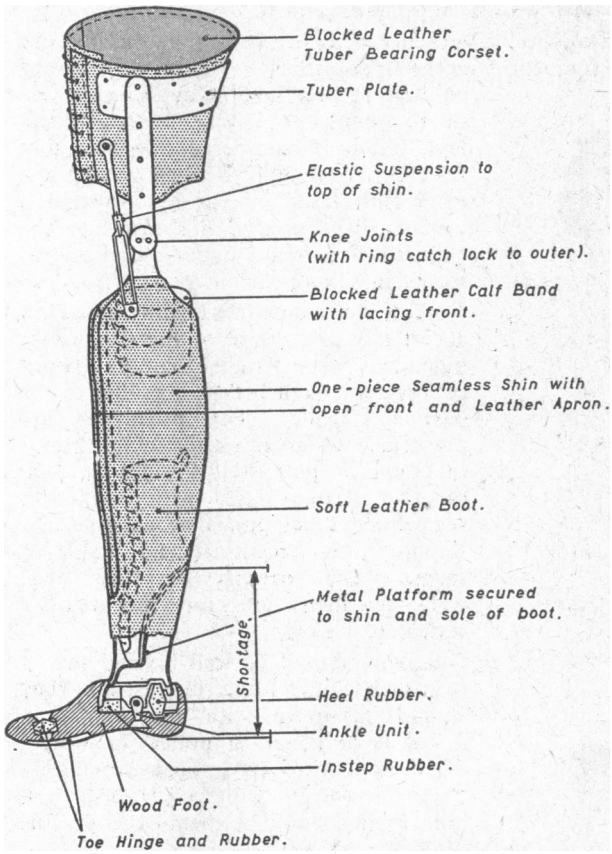

FIG. 4.—An extension limb showing the general principle of concealing a short, deformed limb. It can be modified to the particular deformity. Drawn by Miss Patricia Archer, of Queen Mary's Hospital, Roehampton. Reproduced by permission of the Editor of *Annals of the Royal College of Surgeons of England*; and from *Artificial Limbs* by permission of Pitman Medical Publishing Co. Ltd.

In the upper extremity growth is a relatively minor problem. A young above-elbow amputee may at some time during his growth period require a revision of the conical stump. In a below-elbow stump, where the main growth centre has been removed, this is less commonly required. In the lower extremity, in an above-knee amputation (carried out in a small child at a level comparable to that in an adult) the overall growth of the stump would be reduced by about four-fifths, and therefore the optimal length would never be attained. Hence the preference of disarticulation through the knee-joint. The far commoner occurrence, however, is excessive growth of the bone of the stump in relation to that of the soft parts. This leads first to the "conical" stump and, if unchecked, sometimes to the formation of bursae and ulceration. The discrepancy in rate of growth centre lies proximal to the section—that is, upper arm and lower leg. An amputation stump grows more slowly than the normal rate of growth, similar to the retarded growth of a paralysed limb. Finally, growth of a stump may become uneven, resulting in bowing of the knee stump. This can be observed in forearm and below-knee stumps fitted with prostheses. The cause is obscure. The mechanical effect of the prostheses should not differ greatly from that of a natural hand or foot.

The surgeon should examine the growing stump at regular intervals. It may need revision or epiphysiodesis, or the socket of the artificial limb may require modification or lengthening, because a short prosthesis may result in a pelvic tilt and scoliosis. The whole mechanical structure of the child will function to greater advantage if the length and pressure points of the prosthesis are correct. Some intermittent pressure on the growth centre is of great importance to avoid retardation of growth in children with artificial limbs. This is best achieved by giving child amputees end-bearing stumps. By "end-bearing" I mean exerting direct upward pressure on the end of the bone, preferably after joint disarticulation. This process is essential during the growing period, and the effect can only be seen by observation over a number of years.

When maturity is reached the ordinary type of limb-fitting becomes possible, although it is sometimes necessary to resort to re-amputations at this stage. In badly deformed limbs a disarticulation at the knee or elbow is always preferable to an above-knee or an above-elbow amputation, but if there is a small chance of saving a portion of the limb it is better to perform a below-elbow or below-knee amputation.

Use of Prostheses

It is most important to decide in both upper and lower extremity amputations at what age a child should be provided with an artificial limb. There can be no

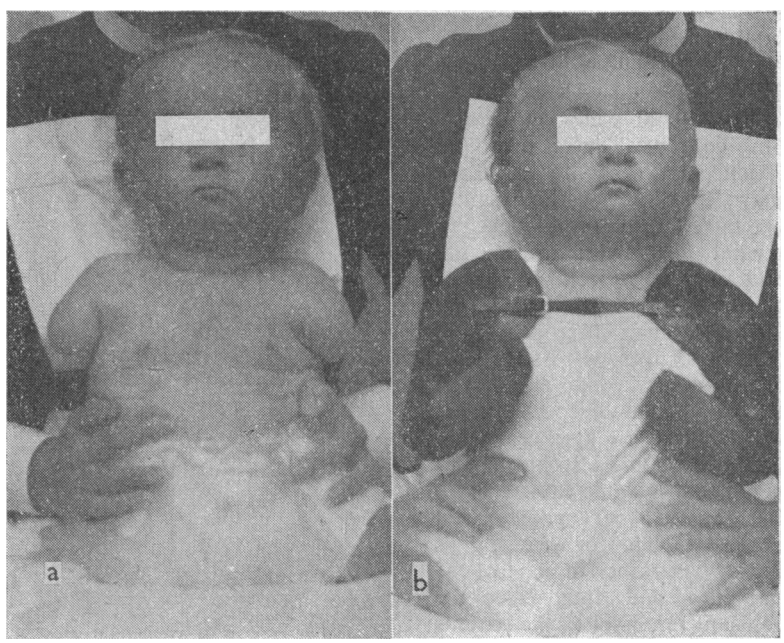

FIG. 5.—Left: An infant with congenital absence of part of both arms, forearms, and hands. Right: Fitted with artificial arms and now able to play and help himself with feeding.

rigid rule, but it can be stated that the child should be fitted as early as possible. The earlier an appropriate appliance is fitted the more opportunity the child will have to adjust himself and become skilled in its use.

Some children have been fitted with arms within the first year of life (Fig. 5). It is important to prevent the child developing a pattern of living with one arm, and

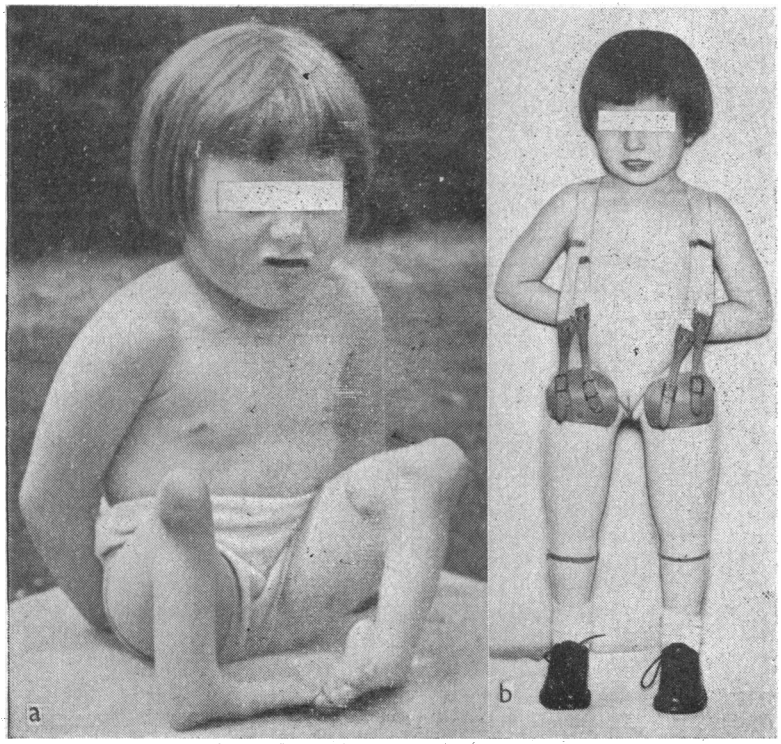

FIG. 6.—(*a*) Child with bilateral congenital absence of tibiae, gross flexion contracture of both knees, and gross deformities of both feet. Disarticulation of both knees. (*b*) Child made fit for school. Reproduced by permission of the Editor of *Annals of the Royal College of Surgeons of England.*

certain instinctive movements of babies have a bimanual pattern—for example, grasping, holding, pulling, pushing, etc. Even in crawling a little artificial arm can be of value.

The age at which an upper-limb congenital defect or amputation stump should be fitted may depend not on the calendar age but rather upon the developmental age, from the mental and physical aspect. While all children should be fitted before attending their first school, it may be dangerous to supply an arm to a very young child. The degree of intelligence present will suggest the time to begin fitting. It should also be remembered that in some cases an arm is fitted to a very young child not so much for the child's benefit but rather to conceal the disability and thus ease the constant reminder to the parents that they have produced an abnormal child. When, however, the young child reaches the age and stage of development when the functional appliances can be fitted the position becomes different. The use of the appliances draws the attention of the parents and others to the disability, and it is the former who need encouragement.

A combined absence of upper and lower limbs is not a frequent congenital deformity. Where such occur, children can be fitted from about the age of 18 months to 2 years with artificial legs, but it is considered advisable that all children should be fitted by the age of 5 with both arm or leg prostheses, because it is around this age that the child starts to make major social adjustments in school and in play.

Arms are sometimes mainly fitted on account of the desire of the parents to hide the deformity rather than on account of any functional benefit accruing to the particular child. Leg prostheses are prescribed as soon as the child begins to crawl or pull itself upright at 8–10 months. Babies fitted with appliances regard the dress appliance as toys and require little encouragement to wear them. The parents at this stage welcome their use for the reasons stated.

Parents should be well briefed that it is in their child's future interest that they should persevere with the appliance. Cases have come to notice in which, when appliances have been supplied—for example, to a little girl now well able to make use of them—the parents discourage their use.

It is well known that children can be unkind to each other, but the presence

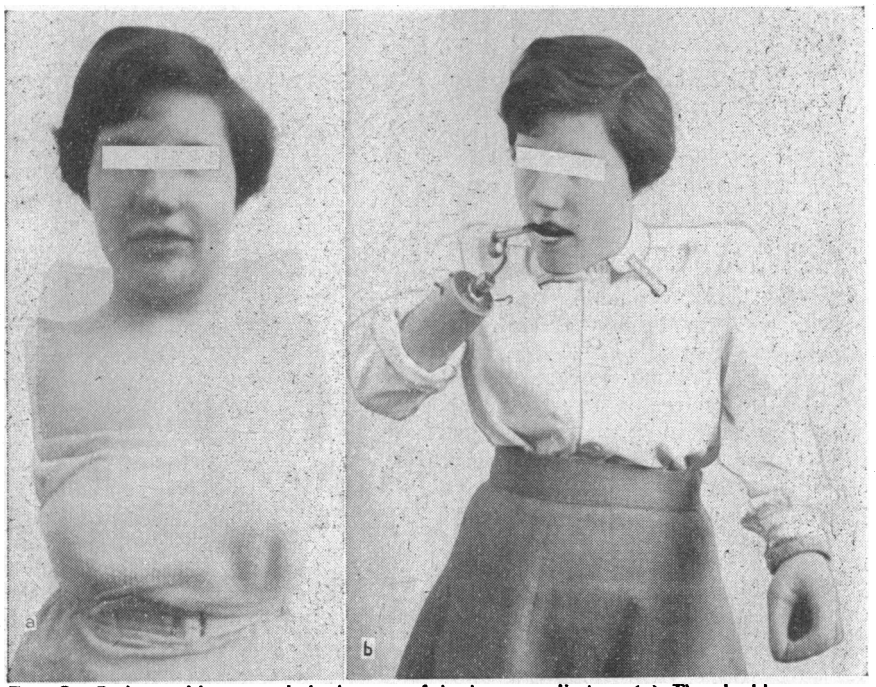

FIG. 7.—Patient with congenital absence of both upper limbs. (*a*) The double exposure photograph illustrates the available excursion of the shoulders achieved by elevation of the shoulder girdles. (*b*) Shows how patient, after being fitted with artificial arms, is capable of doing most of her routine work, including her toilet. Photograph taken by Mr. A. E. Bau, Hugh Steeper Ltd., Queen Mary's Hospital, Roehampton.

of an appliance often lessens the harsh attitude of children to an amputee (Fig. 6 (a) and (b)).

With the object of readjusting these children so that they may lead normal lives they should be regularly inspected and studied to note growth, mental development, changes of talent and interest. Such variables will dictate the relevant alterations in the prostheses that have been fitted, particularly those of bilateral arm amputees.

Children's splints and appliances should be checked by an orthopaedic surgeon or at a limb fitting centre every three months so that growth can be noted and allowed for. Thus compensatory scoliosis or recurrence of deformities can be avoided.

It is important to have a knowledge of all the prostheses available so that appropriate devices can be made to suit a child's special requirements.

Generally speaking, children adjust themselves admirably (Fig. 7, a and b), and take special delight in overcoming difficulties and in learning new manoeuvres with artificial hands.

Besides evaluating the child, the mentality of the parents and other children in the family should also be studied. If the parents lack insight it is imperative to educate them. This is particularly important when dealing with congenitally malformed children. Each parent may consider that the production of a malformed child is due to some defect in the other parent, with resultant domestic disturbances. Others, fortunately few, may tend to neglect such a child, while some are oversolicitous.

Other Possibilities

Among additional lines of treatment are, for instance, kineplastic surgery: the utilization of any residual muscle power by means of skin-lined muscle tunnels. For some children with no arms at all kineplastic surgery remains the last resort if the fitting and training in the use of standard arms, suitably modified, should fail. So far, however, patients fitted with standard arms in this country have succeeded in attaining an efficiency equal to, if not better than, that achieved by kineplasty. But throughout the world intensive research is constantly being undertaken in an attempt to design better and more efficient appliances for these limbless children.

Another line is that of externally powered upper extremity prostheses.

Electric Motors.—Small electric motors installed in hands or elbows and activated by batteries have been used for the activation of upper-extremity prostheses. These have not been widely accepted, since the control mechanism has created many difficulties. Another objection to present-day electrically powered and controlled upper-extremity prostheses is the fact that the movements are slow and robot-like. With increasing knowledge and improved movement devices there is the possibility, however, that electricity may play an increased part in externally powered prostheses.

Compressed Gases.—In Britain and in Germany another method of utilizing external forces has been attempted. Highly compressed carbon dioxide contained in small cylinders is released through valves and is used to extend bellows or pistons which operate the component parts of the prosthesis. Developments along similar lines are being carried out in the United States. The advantages of pneumatically operated prostheses over those which are electrically controlled are (*a*) a more spontaneous action and (*b*) closer control by the amputee.

The main advantage of external power lies in its use for the very severely disabled. The operation of these devices requires frequent replenishment of their power source.

Conclusion

A child born with a defective limb is in a very different position from an older person who has lost a similar part of his body. In the latter, certain movement patterns have developed which can be modified and utilized for the control of an artificial limb. If a functioning limb has never been present the child learns from the beginning to do without it and will develop entirely different movement patterns. This is best seen in those who were born without arms and have learnt to use their feet instead. That is a way of life which certainly deserves admiration, but it is not the one that gives most satisfaction to the person concerned, nor does it offer the greatest scope for developing his potentialities.

It is our endeavour to lead these unfortunate beings to a life as nearly normal as possible by providing them with artificial limbs. The movement patterns required for the control of these limbs are most easily and most perfectly developed at the same time as those for the natural limb. It is for this reason that we attempt to fit a simple and light prosthesis as soon as the child tries to crawl or to pull himself up by his arms. We have, in fact, fitted limbs before the first birthday.

This early fitting is especially important in defects of an upper limb, where the movement patterns are to a far greater extent acquired than in the lower limb where they are largely innate. It becomes the decisive factor in the development of a bimanual pattern of living as opposed to a one-armed one in a unilateral defect.

In practice this aim is best achieved if the surgeon in charge of the case evolves his plan for treatment from the beginning in consultation with a limb-fitting surgeon at one of the Ministry of Health Limb Fitting Centres (*British Medical Journal* (1962)), who can advise not only on the first prosthesis to be supplied but also on the ultimate aims and possibilities.

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Four senior officers retired during the year under the Malayanization scheme; one left on completion of his contract, and another retired on marriage. "These vacancies were filled by appointing the medical officers attached for training to our staff. Our numbers have been restored, but only by replacing experienced senior officers by the young and inexperienced. In itself this is normal, and entirely desirable provided that there are sufficient senior officers left to supervise the work of their juniors. But so many officers have left, or will be leaving within a short time, that there is a grave danger of this Institute being left with almost no senior officers at all. There is no substitute for experience, and two or three years' training can never fit any medical officer, however brilliant, to replace an experienced officer. Some deterioration in the quality of the work at this Institute seems inevitable in the future unless there are sufficient experienced officers to provide adequate supervision." (Institute for Medical Research, Federation of Malaya, *Annual Report 1959*.)