Current Concepts in the Management of the Juvenile Amputee

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Amputations and their prosthetic restorations have, at least in adults, a very long history. Initially wars and then the trauma secondary to a mechanized civilization have produced a large number of amputations in the adult population. Throughout the years an effort has been made to restore these otherwise productive people to a meaningful place in society.

Amputations in children were a relative rarity for many years. As our civilization has become more mechanized, children have been exposed with increasing frequency to the kinds of trauma that may produce loss of extremities (see Table 1). Additionally, there have always been a certain number of children who presented themselves for treatment with a terminal transverse deficiency, sometimes mistakenly called a congenital amputation. The numbers of these patients, even in large general crippled children's services, have been quite small.

In children as in adults, there is very little resistance to the application of lower extremity prostheses. In spite of mechanical inadequacies, most juvenile lower extremity amputees have had some type of fitting early and have continued to use the prosthesis. The upper extremity is an entirely different problem. Historically there was very little in the English literature prior to 1950, concerning age of fitting of upper extremity prostheses and indications for their application in children.

With the tremendous improvement in commercially available components and fabrication techniques of both upper and lower extremity prostheses following World War II, there followed a logical increase in interest in the management of all amputees. As an outgrowth of this, children with amputations were treated more vigorously. There slowly evolved more clear-cut indications for prosthetic restoration, better training techniques, more satisfactory limb fabrication, and eventually well organized prosthetic clinics utilizing the team concept and devoted entirely to the care of children.

As a result of this increased interest in prosthetic restoration, children with congenital limb abnormalities (whose treatment by standard orthopedic reconstructive procedures had been less than ideal) were fitted, on a trial basis, with prostheses. Slowly there developed a new concept in the
management of certain limb deficiencies. At the present time prostheses are utilized in several groups of children:

1) The true post-surgical amputations, regardless of etiology;
2) The terminal transverse limb deficiencies (congenital amputations);
3) Congenital limb deficiencies that do not present themselves as amputations.

The sum of these three groups represents a small but definite portion of the crippled children population. There are, unfortunately, no reliable figures available anywhere concerning the incidence per unit of population of any of these groups.

Prosthetic-Orthotic Education at Northwestern University has instituted as a portion of its regular curriculum a course devoted entirely to
the management of the juvenile amputee. This course, which represents a joint effort of Northwestern University School of Prosthetic Education and the Michigan Crippled Children Commission's Area Child Amputee Center, is specifically concerned with the principles of management of the three types of juvenile amputees that have been mentioned.

**Age of Fitting**

Lower-extremity amputees or children with abnormalities who are to be fitted as lower-extremity amputees should be fitted at the time that the patient is ready to walk, or as soon after surgery as the stump will permit prosthetic application. Normal children develop the ability to stand and walk in an orderly manner. The time sequence is relatively constant within a six months' variation. It is difficult to state categorically that children with lower extremity amputations or limb deficiencies should be fitted at a specific month, but somewhere between twelve and eighteen months of age most children are capable of maintaining satisfactory balance in the erect posture and many have initiated independent bipedal ambulation. When there is observed motor kinesthetic maturation to permit independent standing balance, a lower extremity prosthesis can be applied and ambulation will follow.

In the upper-extremity group, the age of prosthetic fitting may be determined by the goals that are desired from limb application. In the congenital terminal transverse deficiencies, early fitting can be utilized providing all concerned (doctor, patient, family, therapist, etc.) recognize that the prosthesis will not produce a level of function that is greater than the child's potential, i.e., a four-year is not as skillful as a six-year-old.

If one desires skillful prehension functions from an upper-extremity prosthesis, fitting must be delayed until the child has matured to the point that he would be capable of similar functions with a normal upper extremity. In short, then, there are two basic concepts in the fitting of upper extremities: 1) fit as early as the patient is seen, and 2) defer fitting until six to twelve months before the entrance into the public school system (four to four and one-half years of age). Early fitting theoretically accomplishes certain desirable things. It produces equal length of the extremities and thus encourages the child to carry out manual tasks at a normal arm's distance from the body, preventing the development of substitution patterns that bring many of the manual functions to the level of the deformed extremity; prosthetic application masks the sensory function of the stump; the dependence upon visual clues is encouraged; prosthetic tolerance is developed early. For these theoretical but probably valid reasons, early fitting with a passive terminal device—followed by gradual transition to an active terminal device, when the patient's motor skills permit, is recommended.

**Surgery**

The surgery of the juvenile amputee is different from the surgery of the adult amputee in that the juvenile patient has a growing skeletal growth, and therefore a stump can be fashioned at the time of the original surgery which will not change in length because of growth. Amputations in children with immature skeletons must always be fashioned with a full understanding of the longitudinal growth potential that exists in the stump skeleton. A precise knowledge of the contribution of each of the long bone epiphyses is essential. As a general rule, amputations in children are not done at pre-determined sites of election, but the rule of "save all length possible" is recommended. Preservation of epiphyses is imperative; disarticulations, therefore, are more
frequently done in children than are classical supra-epiphyseal amputations. Short stumps, neuromas, scars and spurs are not major complications in children. Tenotomies to make short stumps functional are done much less frequently in children than they are in adults.

The major post-surgical complication in juvenile amputation surgery is "overgrowth." This problem has been recognized for years, and the literature mentions several methods of management. Clinically, overgrowth is an increase in length of the amputated bone with subsequent irritation of the soft tissue, sometimes producing a bursa and in the extreme cases, a perforation of the bursa and skin with secondary superficial infection at the area of perforation. Radiologically the overgrowth presents itself as a sharp spicule of poorly trabeculated bone extending from the end of the stump skeleton. It may or may not perforate the soft tissues. This phenomenon ceases when skeletal maturity is reached. It is seldom seen in congenital terminal transverse deficiencies unless there has been some kind of surgery done on the stump. The relationship of this complication with skeletal immaturity has probably led some authors to believe that this "overgrowth" is related to proximal epiphyseal growth, and epiphysiodesis has been recommended as a treatment. Implantation of metal markers at the tip of the stump skeleton at the time of amputation and subsequent observation by x-ray has demonstrated without doubt that overgrowth is not the result of proximal epiphyseal overgrowth, but represents appositional bone growth from the end of the stump skeleton. In a series of 200 surgical amputations this complication was present in only 8% of the cases. The treatment for this condition is stump revision and removal of the overgrowth. Those patients who develop it once are prone to recurrences. Depending upon the degree of skeletal immaturity when this phenomenon makes its first appearance, a knowledgeable surgeon often can predict whether one or two additional revisions may be necessary. It is believed that this complication does not in itself represent a valid contra-indication to elective amputation in children. Statistically it is insufficient reason to delay an otherwise indicated procedure.

The management of the true post-surgical amputation, the congenital terminal transverse deficiency and the congenital limb deficiency have many similarities, but they are sufficiently different in some aspects so that they should be separated.

Post-Surgical Amputations

Most post-surgical amputations in children are homologues of similar amputations in adults, and in general their prostheses are scaled-down adult models. The fabrication techniques are similar or identical to the adult ones. There are commercially available miniaturized components for both upper and lower extremity prostheses. The principles of fitting and alignment currently taught in adults are applicable in general to children. There are, unfortunately, less accurate check-out procedures for children than there are for adults. This is true both in the upper and lower extremity.

In children with lower extremity prostheses, gait and alignment must be determined on the basis of the child's age and of how children of a similar age walk, rather than by adult standards. In the upper extremity,prehension forces, excursion ranges, maximum openings of terminal devices and percentage calculations of efficiencies of the mechanism must be modified realistically in relationship to the child's age, size, sex and strength. Precise standards have not yet been developed, but experienced therapists and prosthetists are capable of making these modifications very satisfactorily.
Terminal Transverse Deficiencies

Most terminal transverse deficiencies present themselves as homologues of traumatic or post-surgical amputations. The congenital achirexia is a homologue of a wrist disarticulation; the terminal transverse partial hemimelia, upper, is the homologue of a very short below-elbow. Because they are congenital developmental deficiencies, there may be variations in the external configuration of the stump that are not seen in post-surgical amputations. Vestigial remnants of the upper or lower extremity digits, invaginations on the distal end, the presence of redundant skin folds—all are frequently seen. Such external stump abnormalities in this group seldom need any surgical revision. This group in general is readily fitted with standard prostheses using standard components and fabrication techniques. The fit and alignment criteria are similar and sometimes identical to the post-surgical group.

Anomalies

Congenital abnormalities of the longitudinal type in children present bizarre and in many instances nearly indescribable types of deformities. It is very important to recognize that even if the evident defect seems to involve only a portion of the extremity, critical examination will reveal hypoplasia of the remaining mesodermal structures of the remainder of the limb. There are no specific rules concerning the management of these children. Each case is an individual problem and must be evaluated critically as an individual. Such factors as age, sex, family background, rural or urban living, educational facilities and multiplicity of limbs involved all enter into the formulation of a plan of treatment.

The lower-extremity cases of this group generally manifest themselves by leg length discrepancy of a severe degree, malrotation of the limb, inadequate musculature and limited motion in one or more joints.

The upper-extremity group usually manifests itself by arm length discrepancy, alterations in prehension function varying from complete absence to marginal functional ranges of grasp, strength and placement. Here, too inadequate musculature and alteration in range of motion in joints are factors.

In evaluating this group for prosthetic replacement, it is necessary to translate the presenting deformity into an amputation type. From a medical record standpoint, it is desirable that we accurately describe the deformity and give it a name, but from a treatment standpoint we must cease to think of it as a type of deformity and must think of it as an amputation prototype. In order to do this, it is necessary to determine which is the most distal stable joint beyond which there is adequate extremity to function as a stump. Once this level has been determined, then it can be decided whether the presenting abnormality represents a below-knee, above-knee, below-elbow or above-elbow amputation type. The team can then formulate an adequate prescription and with the services of a skilled prosthetist a nonstandard, comfortable socket and prosthesis can be devised. Malrotation can be accommodated for by alterations in alignment of the prosthesis in relationship to the extremity. Nonstandard components are sometimes necessary. Nearly all sockets are nonstandard and of a custom type. Final alignment, fit and gait characteristics in the lower-extremity case must be interpreted in relationship to the specific patient concerned and that patient's particular problem, rather than against a standard.

In the upper-extremity cases, force, excursion and percentage of efficiency must also be modified in relationship to the specific patient, not compared to a standard.
Surgical conversion or reconstruction of upper and lower extremity anomalies in order to fashion more desirable stumps for prosthetic fitting are helpful adjuncts. In a series of 137 lower extremity anomalies, surgical conversion or reconstruction was done in 78 cases (57%). In 108 upper extremity anomalies, surgical conversion was necessary in only 16 cases (15%). It is currently recommended that all upper and lower extremity anomalies be fitted without conversion unless the anomaly is of such a specific nature and with a well enough known life history to establish that conversion is desirable. If at a later date following trial of fitting without conversion it can be prognosticated that conversion will be a benefit, then it should be done.

The problems in this group are further complicated when the patient presenting has more than one limb involved. Decisions then must be made concerning which extremity should be treated first, and next what degree of function should be planned for and whether or not (in the upper extremities particularly) bilateral fitting should be carried out. These are difficult decisions and sometimes can only be resolved on a trial and error basis. It is currently believed that the “multihandicapped” child should not be fitted until sitting balance is established. If the child has both upper- and lower-extremity involvement, the decision as to fitting uppers or lowers first is difficult. Fitting of the lowers improves torso balance, and if the child has one useful upper extremity, the lower extremities should be treated first. If the child has no useful upper extremity, then an attempt should be made to fit for as much upper extremity function as is possible relative to the child’s age and abilities. There is probably no place for the truly inert terminal device for the bilateral upper extremity amelia. Passively operated voluntary-opening terminal devices seem to be indicated even in initial (very early) fittings in this group. Currently it is believed that the bilateral upper amelia should be fitted prosthetically unilaterally, and which side should be fitted can either be decided arbitrarily or, if foot function has developed, fit on the side of the leading foot.

It is currently believed that prosthetic replacement is desirable in children with post-surgical amputations, congenital terminal transverse deficiencies or congenital limb abnormalities of the longitudinal type. Experience has demonstrated that children, if properly fitted and adequately trained, will accept and utilize both upper- and lower-extremity prostheses. In the longitudinal anomalies there is little doubt that prosthetic replacement with or without surgical conversion has in most instances offered more than previous standard reconstructive surgical techniques. Most children with amputations or limb abnormalities treated as amputations have a good rehabilitation potential. With the exception of the “multi-handicapped” group it is believed that these children should be educated in public school systems and they generally demonstrate their ability to compete successfully in spite of their handicaps.