Rehabilitation in Limb Deficiency. 2. The Pediatric Amputee

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This self-directed learning module highlights advances in the evaluation and management of the pediatric amputee. It is part of the chapter on rehabilitation in limb deficiency in the Self-Directed Physiatric Education Program for practitioners and trainees in physical medicine and rehabilitation. This article discusses the etiology and terminology of childhood limb deficiency. Developmental milestones are used as a guide to prescribing prosthetic devices for children, and tables suggesting such guidelines are included. Specific clinical examples are provided to illustrate management issues. Advances that are covered include limb-sparing procedures in the management of lower extremity malignancy.

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2.1 Objective.—Discuss the etiology and terminology of congenital limb deficiency. Develop a management plan for a 5-month-old girl with transverse left upper limb deficiency, forearm, upper third, from presentation to maturity.

Congenital changes are the commonest cause of limb deficiency in children under 10 years of age. Trauma is the commonest cause in the next decade. The cause is unknown in approximately 60% to 70%. Environmental agents such as maternal infections and disease states, amniotic constriction bands, and exposure of the fetus to some known drugs, chemicals, and irradiation are implicated in approximately 10%. About 20% have single gene mutation etiology. Genetic counseling may be indicated in a multi-limb-deficient child or if a child has a known genetic limb deficiency—for example, longitudinal tibial deficiency. Some syndromes, such as TAR (thrombocytopenia absent radii) and some craniofacial disorders are also associated with limb deficiencies. Transverse left forearm, upper third, is the most commonly encountered congenital limb deficiency.

Seventy-five percent of acquired amputations in children are due to trauma from motor vehicles, power tool incidents, gunshot injuries, and high-tension electrical burns; 25% of acquired amputations are due to tumor or disease, such as osteogenic sarcoma, purpura fulminans, and others. Tumor is the most common cause of amputation between the ages of 10 and 20.

The International Society for Prosthetics and Orthotics (ISPO) has developed an international standard based on anatomical and radiological features to describe skeletal limb deficiencies present at birth. Congenital limb deficiencies are divided into transverse or longitudinal. Transverse denotes lack of elements beyond a particular level even though digital buds (nubbins) may exist. Longitudinal denotes reduction (partial) or absence (total) of skeletal elements within the long axis of the limb.

The initial prosthetic fitting of a child with congenital unilateral upper limb deficiency is done at age 3 to 9 months to assist in gross motor development tasks, allowing the use of both upper limbs for creeping, pulling to stand, and so on. Fitting at a later age (2 to 5 years) has been shown to result in greater rejection of the prosthesis because of the development of compensatory techniques. When the prosthesis is first applied, the child may ignore the limb as a result of decreased sensory feedback. Children with acquired amputation may have immediate postsurgical prosthetic fitting or very early prosthetic fitting for optimal use.

In children, the developmental milestones are used as a guide to prescribe a prosthetic device and its components. The components chosen in a child’s prosthesis should provide the needed functional benefit and avoid frustration due to extra weight or difficulty in operation. Passive hands or mitts available in infant size are cosmetic and allow the infants to stabilize their limbs during pull to stand activities. Hook terminal devices (TDs) are available as voluntary opening or voluntary closing in very small and infant size. Hooks provide precision prehension and ease of maintenance. Electric hooks are available in child sizes as switch or myoelectric control.

Body-powered and myoelectric hands are available for children as young as age 2 to 6 years. Body-powered hands provide good cosmesis but weak pinch force and need a harness for suspension and a control cable. Myoelectric hands provide superior pinch force, ability to grip in any position, good cosmesis, easy controls, and less body motion than body-powered prostheses. However, myoelectric components lack durability and cannot be immersed in water. Gloves get torn and soiled and therefore require frequent replacements.

If a child is to receive a myoelectric prosthesis, the passive prosthesis is gradually weighted distally until age 2½ to 3 years. Myoelectric testing is used to assess the voluntary control of the potential muscle groups. In transverse forearm limb deficiency, the wrist flexor and wrist extensor groups are used as potential muscle sites to control the TD. Once the optimal sites have been located, electric toys are used in training the selected muscle groups. In very young limb-deficient children, only one muscle site may be available. In such cases, a single muscle site and function myocontroller, utilizing hand opening and automatic closing, is prescribed.

Children’s wrist units are available as friction wrists that allow passive pronation and supination and serve as the attachment for the TD. Flexion wrist units will allow palmar flexion of TDs and are available in child sizes.
Table 1: Upper Extremity Prosthetic Fitting Guidelines

<table>
<thead>
<tr>
<th>Level of Amputation</th>
<th>Age at Initial Fit</th>
<th>Type of Prosthesis</th>
<th>Milestone (Partial List)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Transradial unilateral: transverse forearm, upper third, unilateral</td>
<td>3-9 mo</td>
<td>Light-weight supracondylar self-suspending socket with possible hand or mitt</td>
<td>When sitting balance is achieved and attempts at lateral propping are made</td>
</tr>
<tr>
<td>Transhumeral unilateral; transverse upper arm, any level, unilateral</td>
<td>6-9 mo</td>
<td>Transhumeral socket, fixed elbow, and passive terminal device (hand mitt or a hook)</td>
<td>Same as above</td>
</tr>
</tbody>
</table>

**Activation**

<table>
<thead>
<tr>
<th>Terminal device</th>
<th>18-24 mo</th>
<th>Myoelectric or body-powered for terminal device</th>
<th>Requires ability to follow simple 2-step commands; interest in bimanual prehension activities. Has reasonable attention span. Gripping in fingers, rather than the fist.</th>
</tr>
</thead>
<tbody>
<tr>
<td>Elbow</td>
<td>36-48 mo</td>
<td>Lock and unlock elbow cable operated or switch control</td>
<td>Understands relationship between objects and immediate environment. Uses prosthesis for bimanual activities as an assisting limb</td>
</tr>
</tbody>
</table>

The modular NYU child-size electric elbow can be used interchangeably with the Hosmer body-powered elbow to operate on a rechargeable battery. The Variety Village Electric Elbow (VV38) can be operated by a pull strap harness, push button, or myoelectric control system and is available for use in children under age 9. However, increased weight of the prosthesis with an electric elbow alone, or in combination with an electric TD, limits applicability in young children. The Utah myoelectric elbow is not appropriate under age 14. Passive friction-control shoulder joints are available in child sizes.

Children have special needs of growth, skeletal maturity, acquisition of new skills, and psychosocial issues dealing with parents, peers, and society. Children with congenital limb deficiency have no sense of loss and consider the prosthesis as an assistive device and not a replacement. If the prosthesis truly works as an assist, then it is accepted; otherwise it is rejected. Prosthetic prescription is not a static phenomenon but changes according to the child’s needs. Attitudes regarding the prosthesis may vary according to the child’s development.

Erickson has provided a framework for recognizing dominant psychosocial issues in different age groups. Infants from birth through age 2 establish trust and security through a very close relationship with their parents. They learn about the world through sensorimotor experiences by touching, pulling things, crawling and reaching, and so on. They compensate for the lost upper limb by using their feet or trunk. As toddlers, they begin to develop a sense of self-worth and independence. School-age children have to deal with peer curiosity and teasing. Adolescents form their own identity and values regarding body image, sexuality, and career. Peer acceptance and cosmesis are a very high priority at this age. Children and adolescents with acquired or congenital limb deficiency may have to contend with a number of psychological and medical stressors. The degree of limb loss has not been found to be a predictor of depression or anxiety in these children. Marital discord in the parents and paternal depression have been noted to predict higher depression and anxiety and lower self-esteem in amputee children. Family support, classmates, teachers, friends, and social support were found to have positive effects on child adaptation.

Studies of cognitive functioning in children with limb deficiencies have so far remained inconclusive. Mild forms of limb deficiency have minimal or no impact on cognitive development. However, according to McDonnell, problems

Table 2: Lower Extremity Prosthetic Fitting Guidelines

<table>
<thead>
<tr>
<th>Lower Limb</th>
<th>Age at Initial Fit</th>
<th>Type of Prosthesis</th>
<th>Milestone/Rationale</th>
</tr>
</thead>
<tbody>
<tr>
<td>Transtibial unilateral</td>
<td>6-9 mo</td>
<td>PTR socket with supracondylar cuff, elastic strap, and waist belt</td>
<td>Child starts pull to stand. Fitting assists in developmental progression to independent ambulation</td>
</tr>
<tr>
<td>Transfemoral unilateral</td>
<td>6-9 mo</td>
<td>Narrow mediolateral socket with no knee joint and a Silesian belt</td>
<td>Same as above. (Child cannot manage a knee joint.)</td>
</tr>
<tr>
<td>Articulated knee joint</td>
<td>Preschool, 36-48 mo</td>
<td>Single-axis knee with elastic extension aid</td>
<td>Can learn to walk with reciprocal gait; extension aid will assist in knee stability.</td>
</tr>
<tr>
<td>Bilateral transfemoral</td>
<td>9-12 mo</td>
<td>Short stubbies with rocker bottom</td>
<td>Can start pulling to stand and take a few steps.</td>
</tr>
</tbody>
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may exist but be difficult to detect. Children with congenital right upper limb defects are more likely to encounter learning difficulties and reading problems than those with left upper limb deficiencies.11 Perceptual motor and cognitive evaluation should be done in all upper limb-deficient children.

Children with acquired amputation have a definite sense of loss and may need a variable period of adjustment.12 Parents in both groups, however, may go through different stages of universal grief response.13 Children pick up the attitudes of others, especially their parents, regarding their body image, self-worth, and attitudes toward a prosthesis. Therefore, it is very important for the parents to become educated and involved in the management of a limb-deficient child as early as possible. Support groups can be helpful in providing a new amputee and family with emotional support, acceptance, understanding, and opportunity to interact with others who are in similar circumstances. Pediatric amputees have been found to achieve generally good functional outcomes.

Play intervention utilizing dolls has been shown to help children in coping with acquired amputation.14 Amputee dolls have proven to be a beneficial instructional aid when professionals are explaining the prosthetic treatment plan.15

The child mentioned in the objective should be fitted as soon as possible with a light-weight below-elbow prosthetic device with a self-suspending or supracondylar socket, friction wrist, and a passive TD. The TD can be a hook, hand, or mitt, depending on the parent's informed choice. A passive hand (mitt) has no grasping function, whereas a hook or a Child Amputee Prosthetic Project (CAPP) TD can hold objects. When the child is 18 to 24 months old, the TD can be activated with the addition of a cable control strap and harness.6

Children should be followed in the juvenile amputee clinic on a periodic basis, especially during growth spurts, until they have achieved maturity. This ensures optimal fit during growth periods and allows substitution with the most appropriate components to maximize function.

2.2 Objective.—Devise a management plan for an 8-year-old boy with traumatic transtibial amputation with a recently developed painful bursa over the distal residual limb. Compare the management of an acquired transtibial amputee with that of a congenital limb deficient child, transverse leg, upper third.

Clinically, the earliest sign of bony overgrowth is a bursa that appears over the distal end of the residual limb and becomes larger and painful as the spur grows. Overlying skin may become reddened and tender as a result of pressure. Painful bursa in children is most commonly due to overgrowth but may also be present as a result of other causes such as poor prosthetic fit. A bony overgrowth is a major complication in congenital and juvenile amputees with acquired amputations under age 17. It does not occur with disarticulation surgery, but it frequently follows metaplastic or diaphyseal amputations, with an incidence of approximately 10% to 30%. Overgrowth occurs as a result of terminal appositional bony growth from the remaining diaphysis. Once the child achieves skeletal maturity, the problem of bony overgrowth ceases. Common sites are the humerus, fibula, tibia, and femur.

Socket modification may delay surgical revision, but once skin penetration becomes imminent or prosthetic accommodation is no longer possible, surgical revision is necessary. Multiple surgical revisions for bony overgrowth are common until skeletal maturity. Surgical residual limb capping using autologous cartilage bone transplant is successful in preventing recurrent bone overgrowth.16 Primary autogenous epiphysseal transplants taken from the amputated limb during amputation surgery to cap the open medullary canal of the residual limb prevents bony overgrowth in acquired juvenile amputations.17

A hand-molded polyethylene foam foot or a commercially available foot with an external keel and a smooth distal border can be fitted to 4-month-old infants. Solid ankle cushion heel (SACH) feet with internal keel and molded toes are most commonly prescribed. They are available in various lengths, are inexpensive, and are durable and light weight. Single-axis feet, though available for children and providing a stable base, need more repairs, and one study showed no difference in the performance of a SACH versus a single-axis foot.18 Dynamic-response feet (energy-storing feet) with flexible keels are available for school-age children and teens. Spring Lite and Seattle Child’s Play are examples.

Flex foot and Flex walk with split toe are available for preteens and teens and allow more spring during jumping and running. Toddlers walk with a wide base of support and external hip rotation, which persist until about age 3. They also walk with excessive knee and hip flexion during stance. The prosthetic socket is aligned in greater abduction and flexion, as a child does not have bimalleolar knee flexion/extension movement until age 2.19

Children cannot control an articulated knee joint until age 3 or 4. Knee joints with a manual lock may be used initially to allow stability, and the lock can be articulated once the child is capable. A single-axis constant-friction knee with an extension aid is durable and inexpensive and is most frequently used. However, it does not allow change of cadence, and the extension aid may need frequent repairs because of wear and tear. Polycentric knees are intrinsically stable and are especially useful for knee disarticulation levels. Pneumatic and hydraulic knee mechanisms are preferred for the active preteen and teenagers because these mechanisms adapt to changes in cadence. They are heavy, expensive, and need more repairs.20

Children, because of their soft, fleshy residual limbs, usually need an auxiliary suspension such as a Silesian band to retain the prosthesis. Total-suction suspension can be used in children by 6 years of age.21

In children, limbs grow faster longitudinally than circumferentially, with the result that children outgrow their prostheses. A young child may start to cry or refuse to wear the prosthesis because of discomfort. Other indications to suggest that a child has outgrown the prosthesis are skin irritation, increase in distal end bearing, residual limb not fitting inside the socket, shoulders uneven during ambulation, gradual limping, or antalgic gait. Some prosthetic options to anticipate growth include use of removable growth liner or five-ply socks, adjustable belts/straps, ½-inch heel lift.
on the contralateral shoe, modular components whenever possible, and extending trimlines. Children usually require a new lower limb prosthesis annually up to age 5, every other year from ages 5 to 12, then once every 3 years up to age 21. They may require frequent interim socket changes and revisions to accommodate residual limb volume changes or any bony overgrowth problems.

Residual limb length is of great importance for the acquired amputee for optimal prosthetic function. If a long transfemoral amputation is performed in a young child, the child will have a very small residual limb, as 70% of the growth of the femur comes from the distal femoral epiphysis. In a child, disarticulation levels are preferred to allow normal bone growth and eliminate bony overgrowth. Some congenital skeletal anomalies may require surgical correction to improve function. In congenital longitudinal deficiency of the fibula, total, there is a severe inequality in leg length. Early foot ablation and prosthetic fitting is the treatment of choice. In congenital longitudinal deficiency of the tibia, total, centralization of the fibula is usually unsuccessful. Early disarticulation of the knee with prosthetic fitting is the treatment of choice.21

2.3 Objective.—Project the functional outcome for a 16-year-old girl recently diagnosed as having osteogenic sarcoma of the distal femur, comparing limb salvage with prosthetic management.

In the past, treatment of osteogenic sarcoma was by amputation; a higher percentage of patients are currently being treated by tumor resection and limb salvage. Reconstruction in such cases includes replacing segmental bone and joint loss by using metallic prosthetic implants, arthrodesis, rotationplasties, and bone allografts.

A modified Van Nes rotationplasty has been used as a limb-sparing procedure since 1981, converting a transfemoral amputation (usually performed for lesions involving the distal femur) into a “functional” transfibial amputation. In the skeletal immature child, careful preoperative planning prior to the Van Nes procedure will allow normal thigh lengths at skeletal maturity. Recent studies have shown no local tumor recurrence, neurovascular compromise, or delay in healing in patients with this procedure.25 Such children are fitted with a modified transfibial prosthesis with a thigh corset, adjustable external knee joints, and a SACH foot. These children and adolescents can approach the activity level of a transtibial amputee.

**References**


*Key References.*

**Resource List—1**

The Amputee Coalition of America, 434 Main Street, Chatham, NJ 07928

Consumer Liaison, American Academy of Orthotists & Prosthetists, 1650 King Street, 5th Floor, Alexandria, VA 22314

National Handicapped Sports (NHS), 4405 East-West Highway, Suite 603, Bethesda, MD 20814

Northwestern University, Rehabilitation Engineering Program, Resource Unit for Information and Education, 345 East Superior Street, Room 1441, Chicago, IL 60611

The Area Child Amputee Center, 234 Wealthy S.E., Grand Rapids, MI 49503

The Association of Children’s Prosthetic Orthotic Clinics (ACPOC), 6300 North River Road, Rosemont, IL 60018


Resource List
