CHAPTER 30

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The Child with a Limb Deficiency

DEFINITION

Generally speaking, limb deficiency is defined as the loss of any part of a limb. This can vary widely in severity, from the unilateral partial loss of a toe as can be seen in constriction band syndrome to the total loss of multiple extremities due to teratogens or genetic syndromes. Limb deficiency can be congenital or acquired. Congenital deficiency may be caused by factors such as genetic syndromes or amniotic bands, while acquired deficiency may be the result of factors such as trauma, severe systemic infection (meningococcemia), or malignant tumor.

Child versus Adult Limb Deficiency. Significant differences exist between the pediatric and adult limb deficiency patient. Children more often have congenital deficiencies, multiple limb deficiencies, and upper extremity deficiencies. Comorbid conditions such as diabetes that are often present in adult dysvascular amputation patients are usually absent in children. Because children are growing, they undergo length and volume changes in their residual limb, and may need more frequent prosthetic and surgical modifications. Phantom pain, which is common in adults, is not as common in children. Children more readily adapt both physically and psychosocially to their situation, and they often have higher functional demands than their adult counterparts. Just as the dictum "a child is not a small adult" is true in all fields of pediatric specialty care, it is also true in caring for the child with a limb deficiency.

CLASSIFICATION

The attempt to classify congenital anomalies has evolved over time to more precisely describe each patient's particular anomaly. The first descriptions of limb deficiencies used terms to describe specific phenotypes, such as "phocomelia" (phoke = seal) to describe the loss of the arm and forearm with attachment of the wrist and hand to the trunk or "lobster-claw hand" to describe the loss of central digits of the hand. A widely used classification system in the United States for congenital anomalies was devised by Frantz and O'Rahilly (1) (Fig. 30-1). This system differentiates between complete limb absence (amelia) and partial limb involvement. Partial limb involvement can affect roughly half of the limb (hemimelia), foot (podos), hand (cheir), digits (dactylos), or phalanges (phalanx). Deficiencies can be transverse, where the distal part of the extremity is lost and the proximal part is relatively normal; longitudinal, where one side of the limb (either the preaxial, postaxial, or central portion) is affected; and intercalary, where the proximal and distal limb are relatively unaffected with an intervening affected segment (Table 30.1).

An international collaboration by the International Standards Organization (ISO) and the International Society for Prosthetics and Orthotics (ISPO) produced a classification system accepted as the universal language of national organizations that treat these children (2). Many of the concepts of Frantz and O'Rahilly are incorporated into this system, although the Greek word roots have been eliminated. This system also uses the concept of transverse and longitudinal deficiencies. In the transverse deficiencies, the part of the segment at which the limb is missing is named, and the extent within that segment may be stated. Thus, complete limb loss at the midtibial level would be "transverse lower leg mid third." In a longitudinal deficiency, the bone or bones missing are named from proximal to distal and described as "partial" or "total." Therefore, a complete tibial deficiency with a hypoplastic great toe would be "longitudinal tibia complete, ray 1 partial."

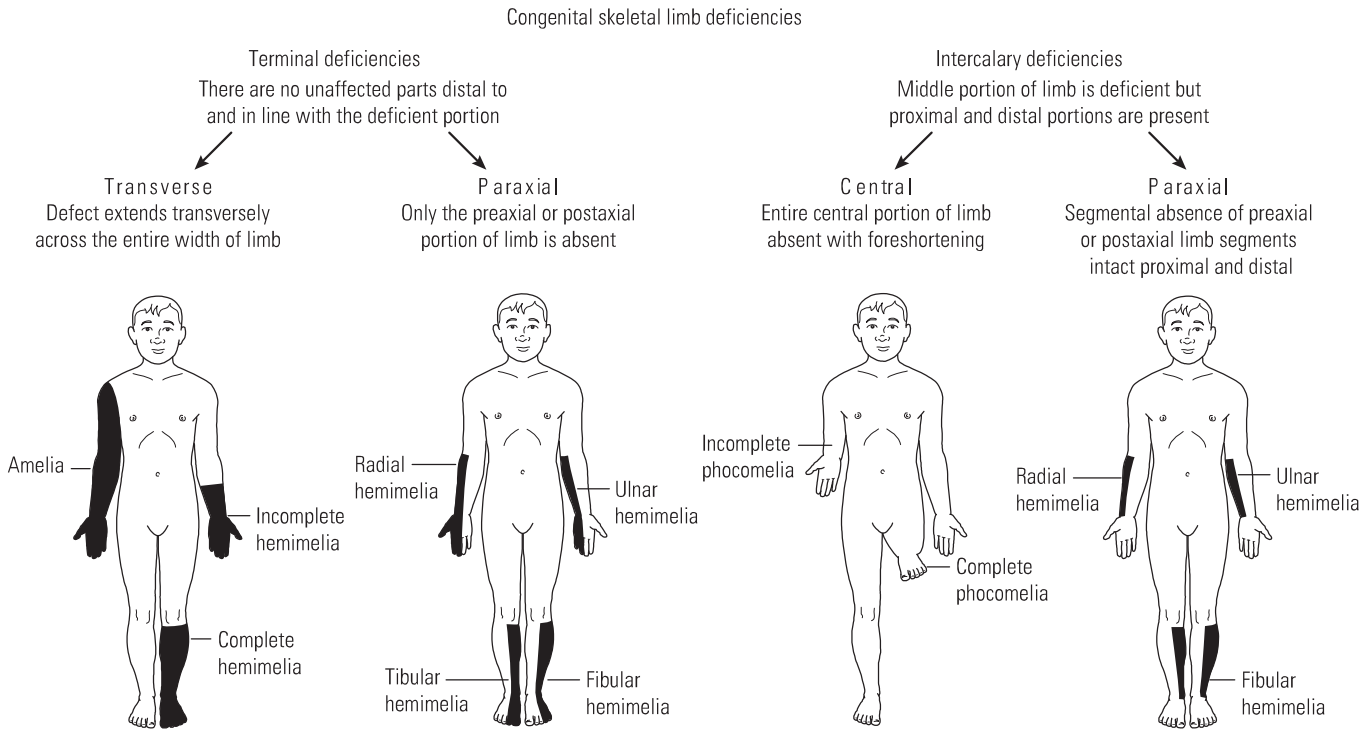


FIGURE 30-1. Diagrammatic representation of the Frantz and O'Rahilly classification of congenital limb deficiencies. (From Frantz C, O'Rahilly R. Congenital skeletal limb deficiencies. *J Bone Joint Surg Am* 1961;43:1202, with permission.)

EPIDEMIOLOGY AND ETIOLOGY

Pediatric limb deficiency is uncommonly encountered by most pediatric orthopaedic surgeons. Depending on which global location is being considered, either congenital or acquired deficiencies may predominate. Congenital deficiencies are more common in developed nations (3), while traumatic amputations can predominate in lesser developed nations (4). Tumors are an uncommon but important cause of limb deficiency in children in all locations. Because limb deficiency is uncommon, patients are often treated in tertiary organized programs that have the experience and resources to treat these patients.

Depending on the methods used, the calculated incidence may vary widely, from 6 per 10,000 in British Columbia (5) to 310 per 10,000 in Tayside, Scotland (6). In a survey of European countries participating in the International Clearing House for Birth Defects Monitoring Programme, the incidence was between 3.1 and 7.9 per 10,000 (7). Statistics such as these are more accurately determined by well-collected birth registries and less accurately determined by surveys from prosthetic clinic medical records, which can overestimate incidence if the clinic is a tertiary referral center.

Fibular deficiency is the most common cause of long bone congenital limb deficiency, when considering that fibular deficiency often accompanies femoral deficiency. Femoral deficiencies are the next-most common, with an incidence between 1 in 50,000 and 1 in 200,000 live births. Femoral deficiencies include the spectrum of the congenital short femur with a stable hip joint and a knee without significant contracture to proximal femoral focal deficiency (PFFD). The prevalence of tibial deficiencies is far less than either fibular or femoral deficiencies and is reported to be approximately one per million live births.

The incidence of all upper extremity amputations is not precisely known but is thought to be more than lower extremity amputations and more often congenital and bilateral than acquired (8, 9). The most common congenital upper extremity amputation is by far the transverse forearm (below-elbow) amputation, with radial longitudinal deficiency being the next-most common. In reality, few pediatric orthopaedic surgeons, other than those working in a limb-deficiency program, will have much experience with these amputations. Although the physician should strive to understand the cause of a congenital amputation in all cases, most of the time no identifiable cause exists. Limb deficiencies can be caused in several ways, such as by environmental factors, genetic disorders, vascular anomalies (such as "the subclavian artery supply disruption sequence"), (10) and amniotic bands.

The oldest and most commonly held etiology for congenital amputation in the past was the mechanical amputation of limbs by amniotic bands, or Streeter dysplasia. Streeter postulated that the bands caused an intrinsic defect in the growth of the fetal limb (11). There is, however, evidence that amniotic bands can form a constriction around the developing limb that interferes with the growth of the limb. The resulting constriction can lead to any degree of damage, from a constriction band around a limb that is otherwise normal to a complete

TABLE 30.1 Classification of Congenital Skeletal Limb Deficiencies

Terminal (T)	
Transverse (-)	**Longitudinal (/)**
1. Amelia (absence of limb)	1. Complete paraxial hemimelia (complete absence of one of the forearm or leg elements, and of the corresponding portion of the hand or foot)—R, U, TI, or FI[a]
2. Hemimelia (absence of forearm and hand or leg and foot)	2. Incomplete paraxial hemimelia (similar to the above, but part of the defective element is present)—r, u, ti, or fi[a]
3. Partial hemimelia (part of forearm or leg is present)	3. Partial adactylia (absence of one to four digits and their metacarpals or metatarsals): 1, 2, 3, 4, or 5
4. Acheiria or apodia (absence of hand or foot)	4. Partial aphalangia (absence of one or more phalanges from one to four digits): 1, 2, 3, 4, or 5
5. Complete adactylia (absence of all five digits and their metacarpals or metatarsals)	
6. Complete aphalangia (absence of one or more phalanges from all five digits)	
Intercalary (I)	
Transverse (-)	**Longitudinal (/)**
1. Complete phocomelia (hand or foot attached directly to trunk)	1. Complete paraxial hemimelia (similar to corresponding terminal defect but hand or foot is more or less complete)—R, U, TI, or FI[a]
2. Proximal phocomelia (hand and forearm, or foot and leg, attached directly to trunk)	2. Incomplete paraxial hemimelia (similar to corresponding terminal defect but hand or foot is more or less complete)—r, u, ti, or fi[a]
3. Distal phocomelia (hand or foot attached directly to arm or thigh)	3. Partial adactylia (absence of all or part of a metacarpal or metatarsal): 1 or 5
	4. Partial aphalangia (absence of proximal or middle phalanx, or both, from one or more digits):1, 2, 3, 4, or 5

-, transverse; /, longitudinal; 1, 2, 3, 4, or 5 denotes the digital ray involved; FI or fi, fibular; I, intercalary; R or r, radial; T, terminal; TI or ti, tibial; U or u, ulnar.
A line below a numeral denotes upper limb involvement; for example, T-2 represents terminal transverse hemimelia of the upper limb. A line above a numeral denotes lower limb involvement; for example, I-1 represents intercalary transverse complete phocomelia of the lower limb.
[a]In capital letters when the paraxial hemimelia is complete, in small letters when the defect is incomplete.
From Frantz C, O'Rahilly R. Congenital skeletal limb deficiencies. *J Bone Joint Surg Am* 1961;43:1202, with permission.

transverse amputation. The previously developed limb has actually been recovered at the time of birth, indicating the mechanism (12). Evidence suggests that most amniotic bands cause deficiency within the first month postconception, based on the fact that limbs and organs commonly affected together are located in close proximity in the embryonic, but not fetal, stage of development (13). Most children with amniotic band syndrome additionally have either craniofacial abnormalities or other evidence of band formation.

Modern genetics has shown that the development of the limb is a complex phenomenon that requires the precise interaction of a large number of genes and their effects, which are described in Chapter 1 and other review articles (14, 15). Many of the proteins and growth factors that participate in this complex interaction have been elucidated (16). Genetic causes of limb deficiency can include chromosomal abnormalities (trisomy 18 and radial longitudinal deficiency), as well as single-gene defects, which result in deficiencies that closely follow Mendelian genetic transmission patterns (tibial deficiency, cleft hand and foot, radial longitudinal deficiency). Even the so-called sporadic deficiencies are more common in families with a history of similar deficiencies. A recent study from the Medical Birth Registry of Norway showed that children born to a mother with a limb deficiency had a relative risk of 5.6% of having the same defect as the mother (17). This is similar to the relative risk of clubfoot. These facts carry consequences for genetic counseling. Understanding the cause of the deficiency is important to the resolving of the guilt that parents will initially feel. The possibility of a transmissible defect is certainly something both they and their affected offspring will also need to know. For the physician, knowing the existence of medical comorbidities and the natural history of the syndrome is necessary for the care of the child.

The disruption of the subclavian artery and its blood supply to the tissues explains the overlap of many of the common orthopaedic conditions seen, for example, Poland syndrome, Klippel-Feil syndrome, Mobius syndrome, Sprengel deformity, and transverse limb deficiencies. There are several possible

mechanisms by which this disruption may occur. For more details, the reader is referred to an excellent review article (18).

Teratogens are an uncommon cause of limb deficiencies. Although typically thought of as medications, there are several other categories of teratogens during pregnancy, including maternal illnesses, medical diagnostic procedures, or trauma. To establish if a particular exposing agent is a teratogen, it should exhibit a dose–response relationship with the defect in question, and it should exhibit a period of greatest sensitivity. Thalidomide, an antinausea medication used in pregnant women in the 1950s and more recently as a chemotherapy agent, caused typical limb deficiencies during a narrow window maternal exposure (between 40 and 44 days postconception). It remains one of the most well-defined teratogenic agents causing limb deficiency. Other drugs are known to affect limb morphogenesis, such as warfarin, phenytoin, and valproic acid. Phenytoin and misoprostol have been shown to affect the vascularity of a previously normally developed fetal limb (19). Retinoic acid and its related metabolites have an effect on limb bud development and cause a wide range of limb malformations in experimental models, which are described in an excellent review article (16).

Almost all of the potential causes of limb deficiency previously described can and do affect other organ systems, often in recognizable patterns. This is an important fact for the treating physician, who should perform a thorough examination for other abnormalities and any heritable genetic defect should be identified. A knowledge of syndromes with limb deficiencies will help the physician look for associated abnormalities and enlist the help of other relevant medical subspecialists when appropriate.

PSYCHOSOCIAL ASPECTS IN LIMB DEFICIENCY PATIENTS

The Parents. When a child with a congenital deformity or congenital amputation first presents, it is the parents who will need the most attention. Although surgical treatment of the child's deficiency is not urgent, the parent's emotions and expectations of treatment understandably take on an urgent tone. Unless prenatal ultrasound detected the deficiency, their child's situation is unexpected and begins a grief cycle not unlike that described by the Swiss physician, Elisabeth Kubler-Ross (20). It is this dynamic that the physician, who is likely meeting the family for the first time, must negotiate. In general, the physician's responsibility over the next several months is both to suggest treatment according to the best medical standards and to guide the parents through the grief cycle.

Parents will initially feel shock and helplessness, which can manifest as feelings of guilt. What did I take or do during my pregnancy to cause this? In many families, there will be a strong desire to know "why this happened." It is a time of anticipated joy that has turned into a period of great stress for both the individuals and, often, the marriage. The initial patient visit is important in establishing a positive doctor–patient relationship by active listening first, then explaining in clear terms the nature of their child's deficit. Obtaining a genetic consultation, even when the deficiency is not known to be hereditary or caused by teratogens, can be therapeutic in this regard, giving the parents additional reassurance.

After the initial shock, parents will have many questions about their child's future. The parents probably have never known a child or an adult with an amputation and have no frame of reference to answer these questions. Although the physician can give lengthy answers to these questions, it is important not to give them too much information all at once. The best one can do during the first visit of the parents is to gain their confidence and give them realistic hope. Fortunately, there are usually no emergent decisions to be made, and there is time to help the parents answer these questions for themselves.

During this initial period, physicians need to be careful about what they tell the parents. In an effort to help the parents feel better, physicians can be tempted to offer false hope and mention treatments that are totally unrealistic. Physicians who do not know, or who do not wish to take on this role, should assure the parents that they are referring them to the best possible care rather than tell them not to worry and that medical science has amazing cures today.

Parents will often refuse a recommendation such as an amputation (21). It is important for the treating physician to recognize the factors that affect their decision and to do his or her best to educate the parents. From the parents' point of view, their child may have a near-normal-appearing foot and a limb that is only slightly shorter than the normal one. Likely, their child will have little difficulty walking, and the parents may not understand the progressive shortening that will develop with time. Along with these observations, they share the popular public belief that modern science can cure everything—next year, if not now. They have all heard of miraculous lengthening of limbs in the popular press, and more recently, the "successful" transfer of limbs. It can be difficult to align the parents' expectations with reality.

After the initial visit, the next several months are a good time for the parents to meet children with similar deficiencies and to see what their child might actually be like in the future. The literature demonstrates that although parents benefit from the support of friends and health professionals, they do not receive the level of support they need (22). This support is found by contact with other parents whose children have similar disabilities. Meeting families and children in a similar situation is comforting, makes the future seem more manageable, and is one of the most important parts of treatment for these children. In addition, the parents can see the various surgical options that might be recommended for their child. It can help them form realistic treatment expectations by seeing what is possible rather than reading and hearing second-hand from other sources, such as the lay press, Internet, or well-meaning friends. Because there is no need for intervention in the congenital deficiency for a few months to years, the parents have time to learn about their child's problem.

After parents have seen other children with limb deficiency, it is a good time to revisit their child's diagnosis and medical treatment plan. By this stage, parents have a lot of information and notions of treatment, and they need guidance to place their child in the correct context of the information. The relationship between the parents and the physician (as well as all members of the team) is important. In this regard, the first thing the parents must be made to understand is that all the decisions will be theirs. Empowering well-informed parents to seek realistic treatment solutions for their child is an essential part of the treatment process. The physician's role is to educate the parents through repetitive explanation and answering parents' questions to ensure they are indeed informed. It is often the other members of the care team, such as the therapist and prosthetist, that the parents will usually "hear" and retain more information from at this stage. Again, nothing helps like seeing other patients and parents.

Finally, the physician should constantly reinforce that the child with a limb deficiency is more normal than abnormal. During this first year, the parents need to resolve their disappointment and loss, accept the child, and see the potential for the future. As part of that process, they need to bond with the child and begin to view the child as an independent person. Most parents will begin to see their children this way with growth and development. Again, this process is facilitated by seeing other children with similar problems. One recommendation that often needs repeating to the parents is that children tend to acquire the fears of their parents and that supporting any activity the child expresses interest in will allow the child to develop his or her natural abilities to the fullest.

The Child. The child with an amputation is essentially different from other children. Although this is evident at birth in the case of congenital amputation, the child will not understand this difference until much later. Children's understanding of their disability is general and incomplete at 6 years of age, but within a few years, around the age of 8 or 9 years, they come to a much more complete understanding of their handicap (23). Therefore, if parents have not discussed this with the child, they can expect the more difficult questions from their child to begin at around this age.

All children with disabilities are vulnerable to social isolation. This, in turn, can have negative effects on the development of self-esteem, body image, and the child's identity, which are developed through interaction with parents, teachers, friends, classmates, and others. As children develop these interactions, the issue of "first appearance" becomes important because it serves as a clue to perceived personal characteristics and can be an obstacle to further healthy interaction. Children in peer groups tend to devalue those with physical differences, a factor that may greatly interfere with these relationships (24). Parents especially understand this and fear for their child in this regard.

There has been a great deal of study on the unaffected child's reaction to various limb deficiencies, showing that children prefer other unaffected children and that they dislike some limb deficiencies more than they dislike others (25–27). In addition, it is known that young adults show signs of anxiety when face-to-face with a person with a limb deficiency. However, there is some evidence that young children do not share their parents' values toward various limb deficiencies when young, but between the ages of 6 and 18 years, they gradually develop values almost identical to those of their parents (26). This would suggest that these values may be subjected to modification among young children and emphasizes that organized discussions with classmates in school about the child's handicap may be of great value.

Despite the negative "first impression" that physical differences hold for children, there is evidence demonstrating that the age of the patient, the gender, the degree of limb loss, or socioeconomic status are not predictors of low self-esteem or of depressive symptoms. Rather, social factors, for example, stress and hassle, parental discord, and social support from classmates, parents, and teachers, along with the child's own perceptions of competency and adequacy, gained through peer acceptance, scholastic achievement, and athletic accomplishments, play the largest role in the development of self-esteem (28–30).

The importance of this information for parents, physicians, therapists, prosthetists, and teachers is that although limb deficiency is the visible problem and is subject to little modification, the important factors in the development of self-esteem are independent of the deficiency and can be positively affected.

CARE COORDINATION

The management of pediatric limb deficiency is best undertaken in a multidisciplinary specialized clinic. There are several reasons for this. First, limb deficiency is rare, and tertiary limb deficiency centers allow participating caregivers to gain the necessary experience to successfully treat these patients. It is beneficial to have these caregivers in the same place at the same time. This makes it convenient for the parents to receive their child's care and can facilitate the efficient transfer of information, both between treating professionals and parents as well as among different treating professionals. Second, as previously mentioned, all patients and parents benefit by knowing they are not alone. They will benefit immensely by seeing and speaking to other parents and children like themselves.

The team should be made up of a physician and surgeon, a prosthetist, a physical and occupational therapist, and a social worker or child psychologist, all of whom are knowledgeable in normal childhood development and who can anticipate the deviations that will occur in development. As compared to adult acquired amputees, who know what they had and what they want, the child and parents of a congenital amputee know little and need education and guidance that usually cannot be provided by an orthopaedic surgeon referring the patient to a prosthetist. The parents will be making decisions for their child with lifelong consequences, and they are acutely aware of

this responsibility. The professionals caring for the family must provide the necessary education and framework in which the parents can make these decisions.

Family involvement is an essential part of the treatment program (31). The child has a condition he or she will adapt to, rather than a disease that can be cured. Hence, the condition should not be "medicalized," but rather treated within the context of family, home, school, and play, not through clinic visits.

GENERAL TREATMENT PEARLS AND PITFALLS

Predicting Growth. It has been observed that the percentage of shortening in a congenital limb deficiency remains relatively constant. This is sometimes referred to as the "rule of proportionality." This principle has been established for congenital short femur (32–34) and fibular hemimelia (35, 36). Clinical experience indicates this to be true for the tibial hemimelias also. It follows from the rule of proportionality that differences in limb length will increase as the child grows. Therefore, in discussing centimeters of shortening and planning treatment, it is important to calculate what the discrepancy will be at maturity rather than focus on what it measures currently. This can be roughly estimated by knowing only the percentile height of the child. With this information, the length of the femoral and tibial segments of the normal limb can be estimated from the Green and Anderson growth charts (37) (Tables 30.2 and 30.3). Then, knowing the length of the normal segments and the percentage by which the affected segments are short, the length of the affected segments at maturity can be estimated.

Although this method of calculating the eventual discrepancy at maturity is clinically valid, the clinician should be aware of the effect that surgical procedures could have on the growth of the limb. Following amputation, the epiphysis of the bone may not grow at the normal rate. Christi et al. (38) showed that in 20 below-knee (BK) amputations in children, only three tibias grew at the expected rate. The congenital group of tibias grew to 36% of what would have been expected, and the acquired group grew to 53% of the expected level. This may be due to the lack of stress across the growth plate, the decreased blood flow to the bone, or the result of the congenital insult that produced the limb deficiency.

Timing of Amputation. The timing of an amputation in a congenital limb-deficient child is tied into the developmental age of the child. In general, amputations for lower extremity congenital deficiency are elective and designed to aid prosthetic fitting. As such, amputation is best performed a few months before the child is developmentally ready to walk (usually when the child is pulling to stand). This gives enough time for the residual limb swelling to subside and for fabrication of the prosthesis. This will allow the child after surgical recovery to maintain a normal developmental sequence. In rare instances, a deformed extremity can interfere with crawling, and amputation may be performed earlier. However, prosthetic fitting in such children should wait until it will be of some functional value. In other cases (e.g., PFFD), amputation may be done at a slightly later age (and *after* prosthetic fitting because of technical reasons).

Although it is poorly documented, there is the impression among both parents and surgeons that with early amputation the child does not experience the body image loss that accompanies amputation at a later stage. Also important is that as a general rule, the earlier the amputation, the better the adaptation of the child's neurologic plasticity to the alteration. No upper age limit has been identified, although most amputations should be performed before school age, if possible.

Overgrowth. Bony terminal overgrowth at the end of the residual limb is the most common problem in juvenile amputees. Its occurrence is reported to be between 20% and 50% and depends on the cause of the amputation, the age of the patient at the time of amputation, the bone involved, and the location within the bone involved (39–41). It occurs most commonly following traumatic amputation or elective amputation through a bone. It is less often seen in congenital amputations because of amniotic band syndrome but not in those due to failure of limb development. It is not seen in amputations through joints (42). Overgrowth occurs most often in below-knee amputations, with the problem being present in the fibula more often than in the tibia, and in transhumeral amputations. The incidence of overgrowth is less common if the primary amputation is performed before the age of 12 years. Recurrence is common and is felt by some to be more common during periods of rapid growth when bone turnover is high (e.g., adolescence).

Contraction of the soft tissue and physeal-mediated growth of the bone, pushing it through the skin, were originally thought to be responsible for bone overgrowth. Aitken disproved these theories when he demonstrated by implanting metallic markers that the overgrowth took place distal to the end of the bone (39, 43). The new bone is periosteal and endosteal appositional bone. Overgrowth results from the typical process of wound contracture as has been demonstrated by Speer (44). Following a through-bone amputation, the periosteum continues to grow. As it grows over the end of the bone, it grows over the open medullary canal, where it contracts and is drawn into the canal from which it can continue to grow, producing the overgrowth at the end of the bone.

Patients with terminal overgrowth present clinically with pain on weight-bearing or prosthetic use. An antalgic gait with decreased stance time may be noticed. Decreased range of motion, to limit pulling of the skin at the end of the limb, is an additional symptom. Clinically, the patient presents with tenderness and pain at the end of the residual limb. There may be inflammation, bursal formation, or the bone end may be protruding through the skin. Commonly, the bony spike can be palpated within a small, tender bursa (Fig. 30-2A–C).

TABLE 30.2 Girls: Lengths of the Long Bones Including epiphyses[a]

Femur

Number	Age (yr)	Mean	σ_d	σ_m	Distribution			
					$+2\sigma_d$	$+1\sigma_d$	$-1\sigma_d$	$-2\sigma_d$
30	1	14.81	0.673	0.082	16.16	15.48	14.14	13.46
52	2	18.23	0.888	0.109	20.01	19.12	17.34	16.45
63	3	21.29	1.100	0.134	23.49	22.39	20.19	19.09
66	4	23.92	1.339	0.164	26.60	25.26	22.58	21.24
66	5	26.32	1.437	0.176	29.19	27.76	24.88	23.45
66	6	28.52	1.616	0.197	31.75	30.14	26.90	25.29
67	7	30.60	1.827	0.223	34.25	32.43	28.77	26.95
67	8	32.72	1.930	0.236	36.59	34.66	30.78	28.85
67	9	34.71	2.117	0.259	38.94	36.83	32.59	30.48
67	10	36.72	2.300	0.281	41.32	39.02	34.42	32.12
67	11	38.81	2.468	0.302	43.75	41.28	36.34	33.87
67	12	40.74	2.507	0.306	45.75	43.25	38.23	35.73
67	13	42.31	2.428	0.310	47.17	44.74	39.88	37.45
67	14	43.14	2.269	0.277	47.68	45.41	40.87	38.60
67	15	43.47	2.197	0.277	47.86	45.67	41.27	39.08
67	16	43.58	2.193	0.268	47.97	45.77	41.39	39.19
67	17	43.60	2.192	0.268	47.98	45.79	41.41	39.22
67	18	43.63	2.195	0.269	48.02	45.82	41.44	39.24

Tibia

Number	Age (yr)	Mean	σ_d	σ_m	Distribution			
					$+2\sigma_d$	$+1\sigma_d$	$-1\sigma_d$	$-2\sigma_d$
61	1	11.57	0.646	0.082	12.86	12.22	10.92	10.28
67	2	14.51	0.739	0.090	15.99	15.25	13.77	13.03
67	3	16.81	0.893	0.109	18.00	17.70	15.92	15.02
67	4	18.86	1.144	0.140	21.15	20.00	17.72	16.57
67	5	20.77	1.300	0.159	23.37	22.07	19.47	18.17
67	6	22.53	1.458	0.178	25.45	23.99	21.07	19.61
67	7	24.22	1.640	0.200	27.50	25.86	22.58	20.94
67	8	25.89	1.786	0.218	29.46	27.68	24.10	22.32
67	9	27.56	1.993	0.243	31.55	29.55	25.57	23.57
67	10	29.28	2.193	0.259	33.67	31.47	27.09	24.89
67	11	31.00	2.384	0.291	35.77	33.38	28.62	26.23
67	12	32.61	2.424	0.296	37.46	35.03	30.19	27.76
67	13	33.83	2.374	0.290	38.58	36.20	31.46	29.08
67	14	34.43	2.228	0.272	38.89	36.66	32.20	29.97
67	15	34.59	2.173	0.265	38.94	36.76	32.42	30.24
67	16	34.63	2.151	0.263	38.93	36.78	32.48	30.33
67	17	34.65	2.158	0.264	38.97	36.81	32.49	30.33
67	18	34.65	2.161	0.264	38.97	36.81	32.49	30.33

[a]Orthoradiographic measurements from longitudinal series of 67 children.

From Anderson M, Messner MB, Green WT. Distribution of lengths of the normal femur and tibia in children from one to eighteen years of age. *J Bone Joint Surg Am* 1964;46:1197, with permission.

Routine evaluation can detect the problem before it is painful, and surgical correction is fairly straightforward at this point. Prosthetic adjustments may help delay surgical revision, but will seldom be sufficient with significant overgrowth.

Treatment of overgrowth has included implantation of various plastic and metallic devices over the end of the bone (the so-called stump-capping procedures). However, these techniques are not commonly used because the results are no

TABLE 30.3 Boys: Lengths of the Long Bones Including Epiphyses[a]

Femur

					Distribution			
Number	Age (yr)	Mean	σ_d	σ_m	$+2\sigma_d$	$+1\sigma_d$	$-1\sigma_d$	$-2\sigma_d$
21	1	14.48	0.628	0.077	15.74	15.11	13.85	13.22
57	2	18.15	0.874	0.107	19.90	19.02	17.28	16.40
65	3	21.09	1.031	0.126	23.15	22.12	20.06	19.03
66	4	23.65	1.197	0.146	26.04	24.85	22.45	21.26
66	5	25.92	1.342	0.164	28.60	27.26	24.58	23.24
67	6	28.09	1.506	0.184	31.10	29.60	25.58	25.08
67	7	30.25	1.682	0.205	33.61	31.93	28.57	26.89
67	8	32.28	1.807	0.221	35.89	34.09	30.47	28.67
67	9	34.36	1.933	0.236	38.23	36.29	32.43	30.49
67	10	36.29	2.057	0.251	40.40	38.35	34.23	32.18
67	11	38.16	2.237	0.276	42.63	40.40	35.92	33.69
67	12	40.12	2.447	0.299	45.01	42.57	37.67	35.23
67	13	42.17	2.765	0.338	47.70	44.95	39.40	36.64
67	14	44.18	2.809	0.343	49.80	46.99	41.37	38.56
67	15	45.69	2.512	0.307	50.71	48.20	43.19	40.67
67	16	46.66	2.244	0.274	51.15	48.90	44.42	42.17
67	17	47.07	2.051	0.251	51.17	49.12	45.02	42.97
67	18	47.23	1.958	0.239	51.15	49.19	45.27	48.31

Tibia

					Distribution			
Number	Age (yr)	Mean	σ_d	σ_m	$+2\sigma_d$	$+1\sigma_d$	$-1\sigma_d$	$-2\sigma_d$
61	1	11.60	0.620	0.074	12.84	12.22	10.98	10.36
67	2	14.54	0.809	0.099	16.16	15.35	13.73	12.92
67	3	16.79	0.935	0.114	18.66	17.72	15.86	14.92
67	4	18.67	1.091	0.133	20.85	19.76	17.58	16.49
67	5	20.46	1.247	0.152	22.95	21.71	19.21	17.97
67	6	22.12	1.418	0.173	24.96	23.54	20.87	19.46
67	7	23.76	1.632	0.199	27.02	25.39	22.13	20.50
67	8	25.38	1.778	0.217	28.94	27.16	23.60	21.82
67	9	26.99	1.961	0.240	30.91	28.95	25.02	23.06
67	10	28.53	2.113	0.258	32.76	30.64	26.42	24.30
67	11	30.10	2.301	0.281	34.70	32.40	27.80	25.50
67	12	31.75	2.536	0.310	36.82	34.29	29.21	26.68
67	13	33.49	2.833	0.345	39.16	36.32	30.66	27.82
67	14	35.18	2.865	0.350	40.91	38.04	32.32	29.45
67	15	36.38	2.616	0.320	41.61	39.00	33.76	31.15
67	16	37.04	2.412	0.295	41.86	39.45	34.63	32.22
67	17	37.22	2.316	0.283	41.85	39.54	34.00	32.59
67	18	37.29	2.254	0.275	41.80	39.54	35.04	32.78

[a]Orthoradiographic measurements from longitudinal series of 67 children.

From Anderson M, Messner MB, Green WT. Distribution of lengths of the normal femur and tibia in children from one to eighteen years of age. *J Bone Joint Surg Am* 1964;46:1197, with permission.

better than the use of biologic material (45, 46). Marquardt reported in the mid-1970s, in the German literature, on the capping of the bone end with a cartilage–bone graft. More recently, Tenholder et al. (47) reported comparable results with the use of a polytetrafluoroethylene felt pad. Various reports for both acquired and congenital amputees indicate generally favorable results, with most revisions being for technical reasons (45, 46, 48, 49).

When a patient presents with pain and bursa formation, prosthetic modification and other conservative measures

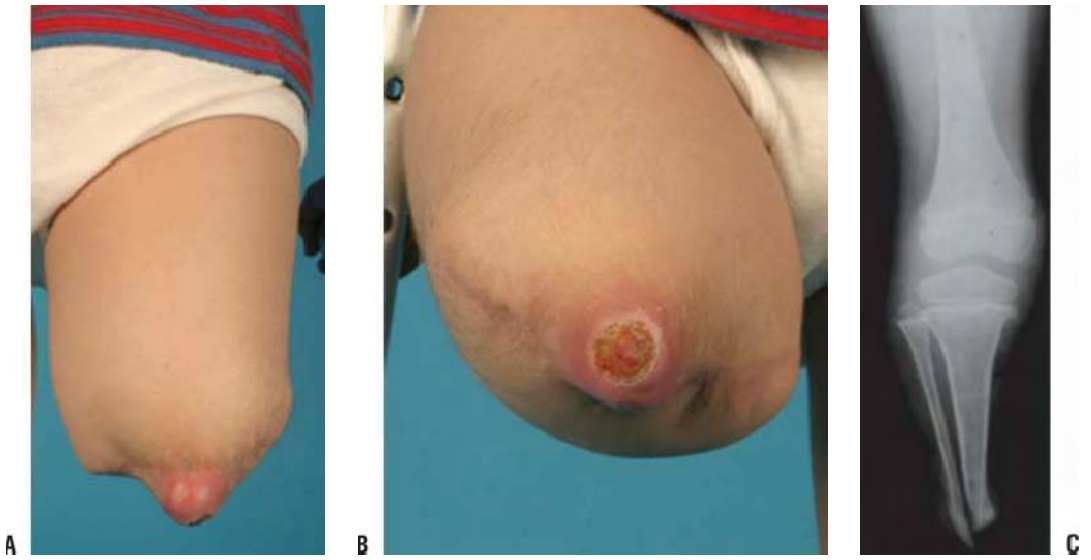
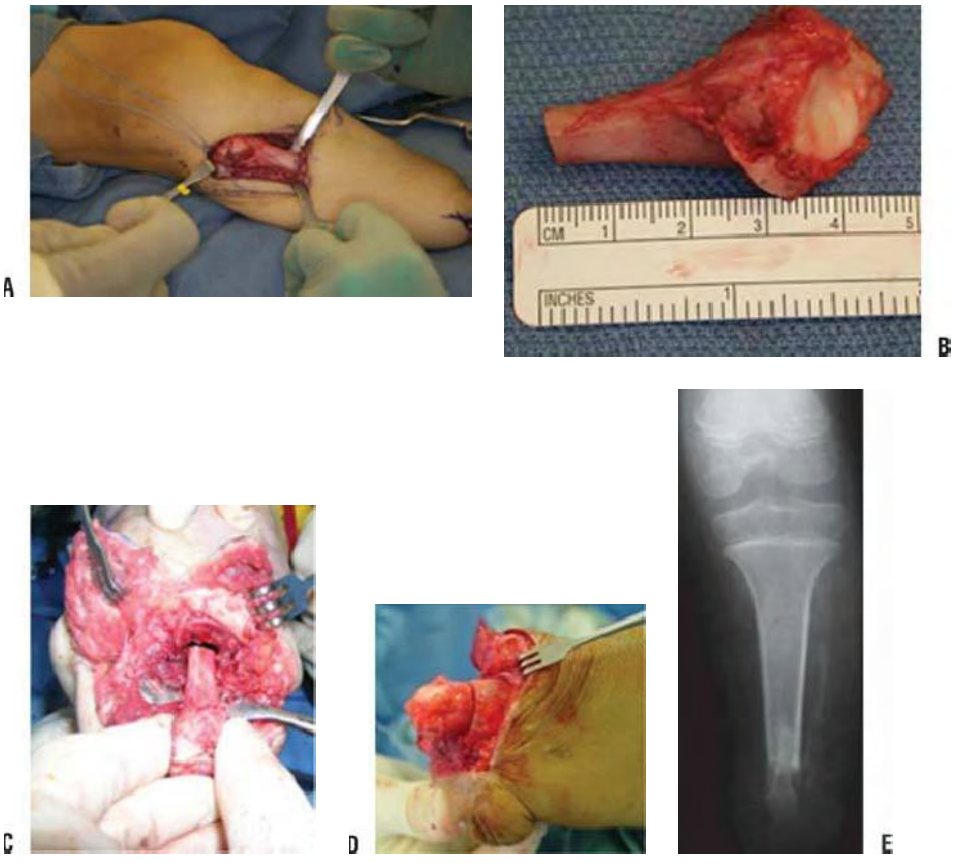


FIGURE 30-2. **A–C**: Clinical photos and AP radiograph of the tibia exhibiting bony overgrowth and ulcer formation.

should be used first unless a bony spike is felt. If revision surgery is necessary, a capping procedure should be considered. In the case of a primary amputation, it is advisable to use available parts from the amputated portion of the limb to cap the end of the bone, if conditions permit. The most common procedure is the use of the proximal fibula to cap the tibia (Figs. 30-3A–E). As in any revision, adequate resection of the bone is essential to provide a healthy soft-tissue envelope. Harvest of the proximal fibula involves detaching the lateral collateral ligament of the knee, which can theoretically lead to lateral knee instability. However, there have been no reports of lateral knee instability after proximal fibular resection for biologic capping, and the literature regarding knee instability after proximal fibular resection for tumors is mixed (50–54). Given that the literature is unclear on the need for lateral ligamentous reconstruction, it seems reasonable to test intraoperative knee instability and repair or reconstruct the ligament if necessary.

FIGURE 30-3. **A–D**: Intraoperative photos demonstrating proximal fibular harvest and subsequent insertion of the proximal fibula in the medullary canal of the tibia (modified Marquardt procedure). **E**: AP radiographic appearance of the tibia 6 weeks after the procedure.



Following surgery, the therapist should supervise and educate the child and parents in edema control to accelerate return to prosthetic use. In addition, range-of-motion and strengthening exercises accelerate, and may even be necessary to regain, the full function of the prosthesis. Users of myoelectric prostheses may need readjustment of their electrodes and, for a while, may have difficulty activating the prosthesis. This is because swelling and reshaping of the limb may alter the optimal sites for electrode placement.

Short Residual Limb. In some patients with either congenital or acquired amputation, the residual limb will be too short for satisfactory or comfortable prosthetic fitting. In such cases, it may be possible to lengthen the residual limb. Watts has written an excellent review of this subject. Favorable results have been reported for both upper and lower extremity residual limb lengthening (55, 56) (Figs. 30-4A–D). Alekberov et al. (57) report on six patients who had successful lengthening of 3.4 to 8.4 cm in congenital below-elbow segments. The remainder of the literature to date consists largely of case reports.

The lengthening of residual limbs is fraught with complications, and careful consideration needs to be given to the potential benefits versus the possible complications. Deficient soft-tissue coverage is the main limit to adequate lengthening. Tissue expanders generally do not provide the solution. Free tissue flaps may be used when skin coverage is inadequate. Free flaps often remove sensation from the end of the residual limb, and especially in the upper limb this can affect the function of the limb both with and without the prosthesis. Although difficult, it is possible to fabricate a temporary prosthesis for use during the lengthening process for both above- and below-knee prostheses so that the child can continue weight bearing, at least during the consolidation phase (55) (Fig. 30-5A–C).

Phantom Sensation/Pain. In the middle of the 19th century, the neurologist Silas Weir Mitchell coined the term "phantom limb." He described sensations as replicas of the lost limb, some being painful and some not. Phantom sensation of the limb is often described by patients as the feeling that they can move the part, tell how the part is positioned, or feel it itching or tingling. Phantom pain, however, is perceived by the patient as just that—painful. It often is the same as the pain before an amputation or may be cramping, shooting, burning, or of any other characterization.

It was generally thought that children born without limbs do not have sensations of them; nor do these children experience the phantom pain or phantom sensation seen in the acquired amputee (58). Recent reports call this commonly accepted truism into question (59, 60). Whatever this pain is, those who care for children with limb deficiencies will recognize that these children do not have the same problem as the true chronic phantom pain seen in adult amputees.

Melzack et al. (60) reported that at least 20% of children with congenital limb deficiency and in 50% of those who underwent amputation before age 6 experienced phantom limb. Of those, 20% of the congenitally deficient group described the sensations as painful, whereas 42% of the acquired amputees described them as painful. To explain the phenomenon of phantom limb in a child who has never had a limb, Melzack et al. have proposed that there is a genetically or innately determined neural network that is distributed in the cortex (not focal), which is responsible for the represen-

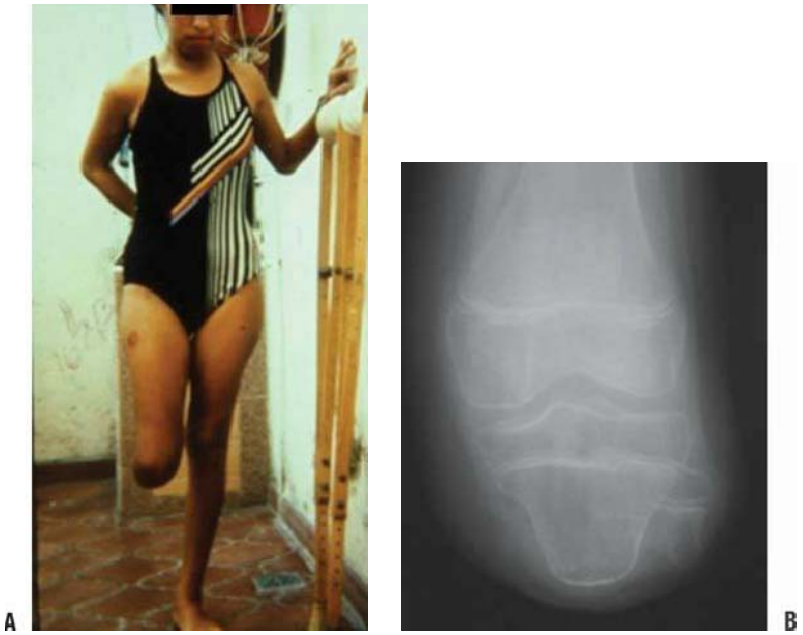


FIGURE 30-4. **A,B**: Preoperative clinical photo and AP radiograph of a patient with a transtibial amputation with short residual limb.

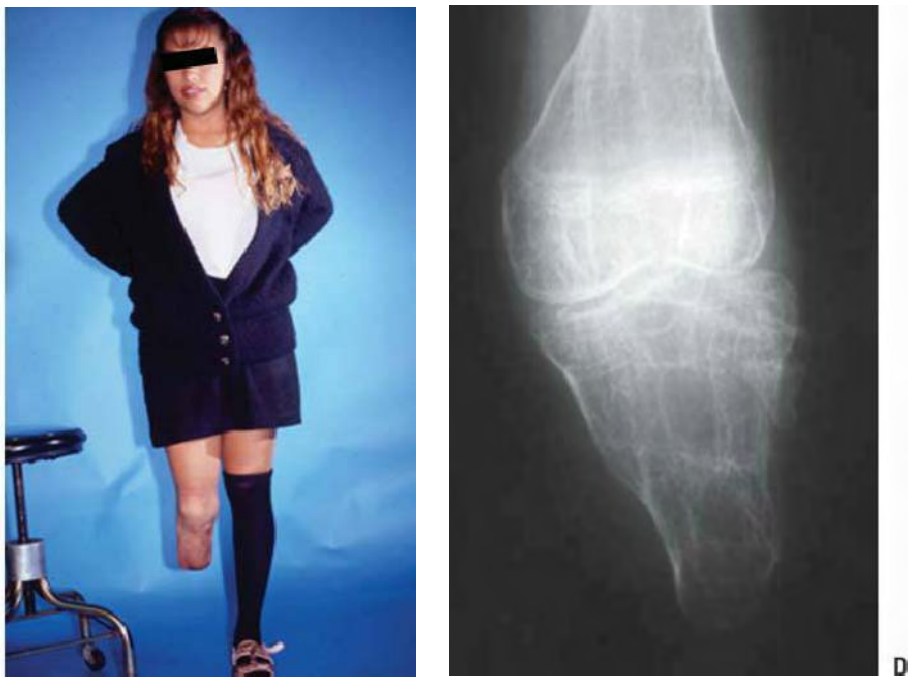


FIGURE 30-4. (*continued*) **C,D**: Postoperative clinical photo and AP radiograph of the same patient after tibial lengthening.

tation of the limb, even in the absence of normal limb bud development.

Phantom pain and distal residual limb pain are also commonly associated with other pains, such as headache, bone, or joint pain (59, 61). The frequency of phantom sensations varies. They are often triggered by a wide variety of stimuli. Feeling nervous or happy, not wearing a prosthesis, being cold, or being ill are frequent triggers. Fortunately, these sensations do not interfere with the child's usual activity, and most children say they just try to ignore the sensations (62).

There is good evidence in adult patients that preemptive analgesia during amputation surgery, or immediately in the postoperative period, can decrease postoperative phantom pain in adults, and it has been suggested that the same is true in children (63). Epidural or spinal anesthesia can decrease postoperative limb pain as compared to general anesthesia (64, 65). Postoperative continuous-infusion intraneural catheters have also been used with success (66, 67). For established phantom limb pain, there is no single highly successful treatment. Because many of these problems resolve with prosthetic alterations or physical therapy modalities, a multidisciplinary approach has proven to be the best intervention in evaluating and properly treating the phantom limb phenomenon when it becomes a problem.

A properly fitting socket, with appropriate suspension and sock thickness, is the best and first treatment of choice (62, 68). A heavy, tight shrinker, either worn inside the prosthesis or when the prosthesis is off, may provide relief. Physical therapy interventions, including weight-bearing and graduating pressures such as tapping, rubbing, and massage to the residual limb, have been reported to give temporary or permanent relief. Rubbing and massaging the uninvolved limb at similar points to those in which they are experiencing the phantom limb sensation may provide relief. Various physical modalities have been utilized in the treatment of phantom sensations in children, including transcutaneous electrical nerve stimulation, biofeedback, ultrasound, and the physical agents of heat and cold (69).

For the occasional adolescent amputee who has problems with phantom pain following an amputation, gabapentin (Neurontin, Park-Davis) has proven a useful medication for some patients (70).

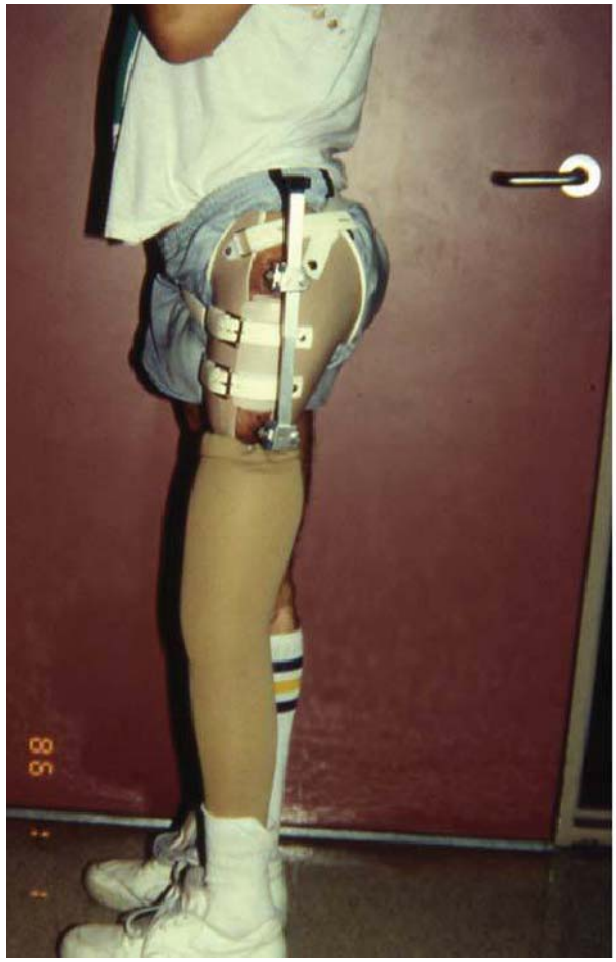
CONGENITAL DEFICIENCIES OF THE LOWER EXTREMITY

Fibular Deficiency

Definition and Classification. According to the ISO terminology, fibular deficiency is a longitudinal deficiency that is either partial or complete. However, this definition does little to accurately portray the spectrum of deficiency that is seen.

Numerous classifications specific for fibular deficiency have been proposed (21, 35, 71–73). To be useful, a classification should guide treatment or aid in prognosis. As treatment changes, it may be reasonable to expect that classifications change. Most classifications are anatomic and are based on the radiographic appearance. Maffulli and Fixsen describe total aplasia of the fibula and a *forme fruste* of the same condition in which the fibula and tibia are short to varying degrees (74, 75). A more specific classification, which is probably the most widely used today, was proposed by Achterman and Kalamchi

A



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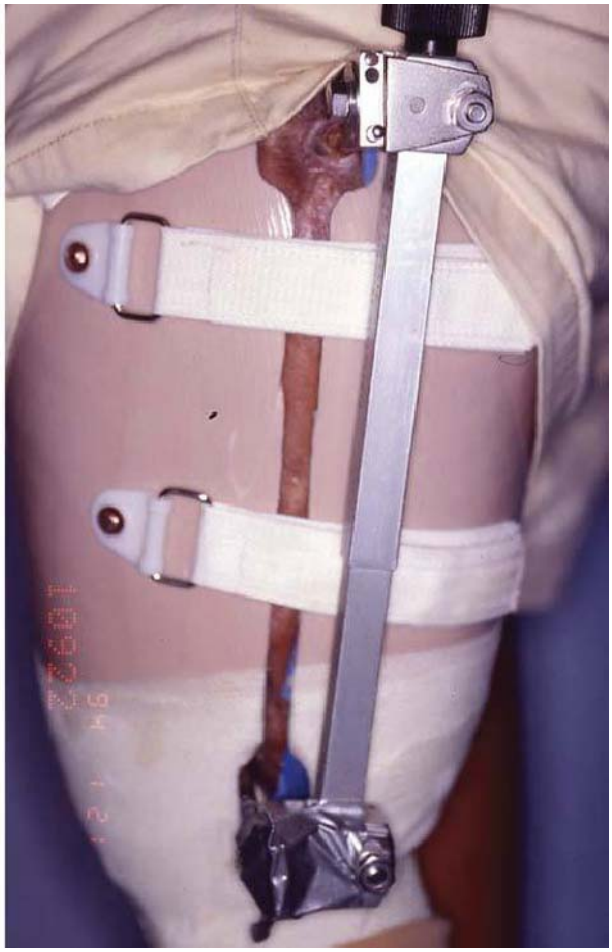
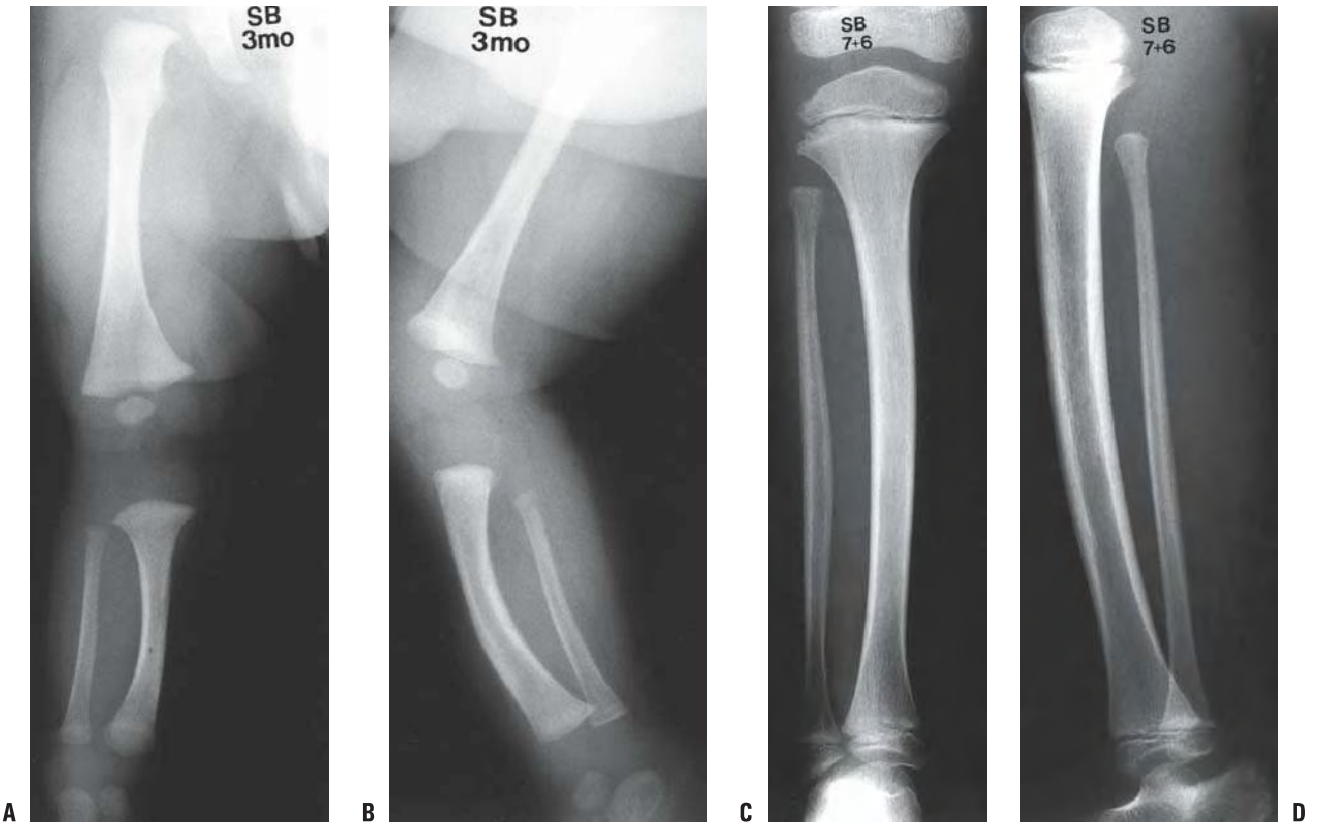


C

FIGURE 30-5. **A–C**: Clinical photos from the front **(A)** and side **(B,C)** of a patient undergoing lengthening of a short residual limb with a congenital above-knee amputation with a monolateral fixator. A modified above-knee prosthesis allows weight-bearing with the fixator in place.

(35) (Figs. 30-6 to 30-8). They correlated the classification with the discrepancy in length and recommended treatment on the basis of the classification. Given the large variation in the different aspects of fibular deficiency, including the parents' desire, it remains unlikely that classifications based on radiographic appearance of the fibula will provide anything more than a rough guide and a method of comparing patients in different reports.



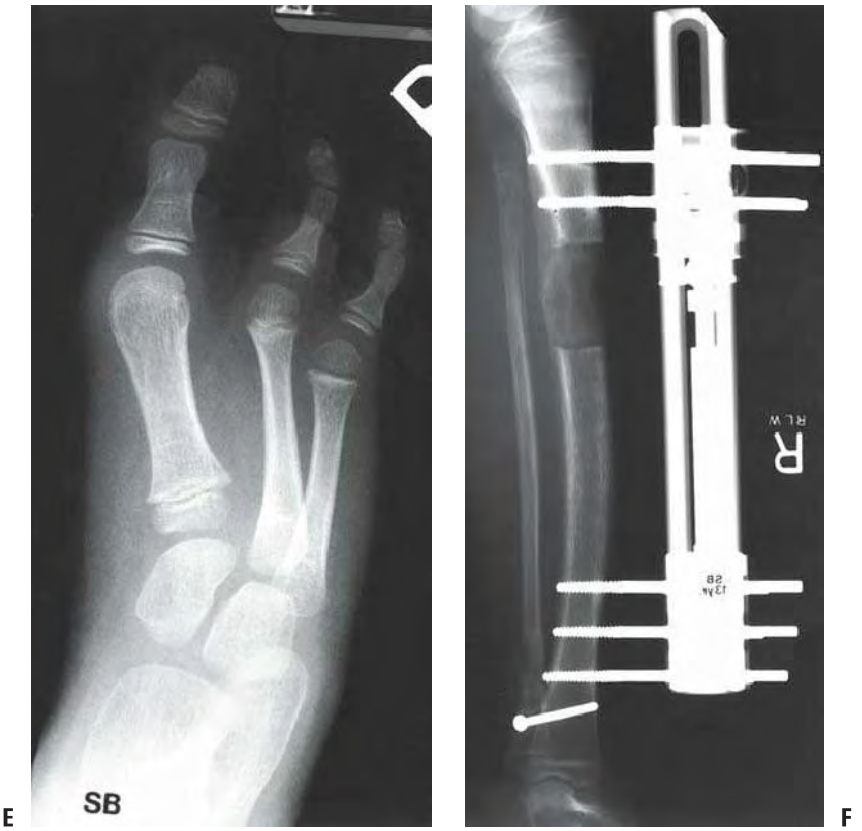


FIGURE 30-6. **A,B**: The radiographs of a 3-month-old boy with type 1a fibular deficiency of the Achterman and Kalamchi classification. The proximal fibula is short. **C,D**: AP and lateral radiographs of the same patient at the age of 7 years, 6 months. The shortening of the fibula is more apparent and the ball-and-socket ankle joint is easily seen. **E**: The foot at the same age, with the lateral two rays missing. **F**: At 13 years of age, the leg was lengthened by 7 cm.

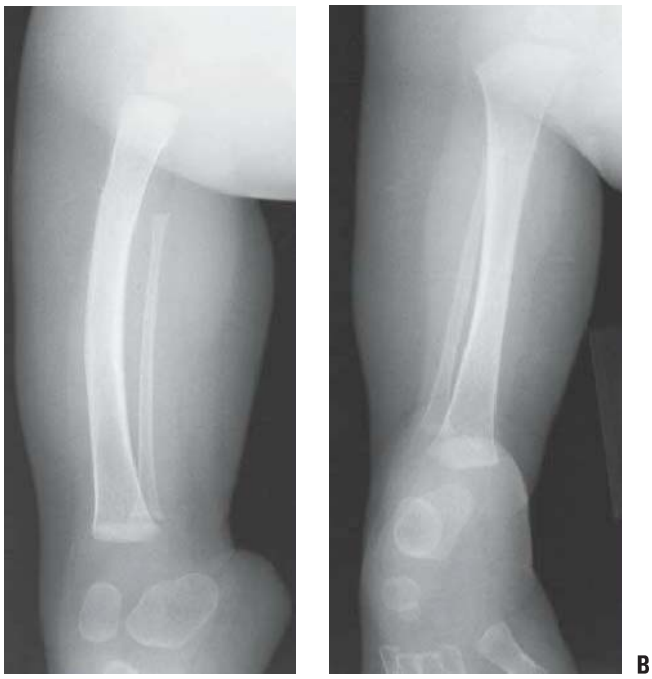


FIGURE 30-7. **A,B**: Type Ib fibular deficiency (Achterman and Kalamchi), in which the proximal fibula is missing. This type is often associated with proximal focal deficiency, as in this child.

Birch et al. (76) have proposed a functional classification on the basis of the functionality of the foot and the limb-length discrepancy as a percentage of the opposite side. The central question in this classification is "is the foot functional?" If it is, the degree of shortening is defined. Those with lesser amounts of shortening can be managed by epiphysiodesis or shoe lift (<5%), single lengthening (6% to 10%), at least two lengthening (10% to 30%), and multiple lengthening or amputation (>30%). For those with a nonfunctional foot, amputation is advised, unless there is concomitant upper extremity deficiency such that the foot is functionally used as a hand.

Epidemiology and Etiology. Considering congenital lower extremity limb deficiencies, fibular deficiency is the most common long-bone deficiency, with an incidence between 7.4 and 20 per million live births (5, 77). The incidence of fibular deficiency would be much higher if up to 80% of patients with PFFD having fibular deficiency were included. The etiology of fibular longitudinal deficiency is not known but is sporadic.

Clinical Features. Although the name fibular deficiency implies a localized deficiency, patients often have typical clinical features throughout the entire limb. There is a wide variation in the scope of the deficiency. Typically, the limb is characterized by a rigid valgus foot, often with one or two

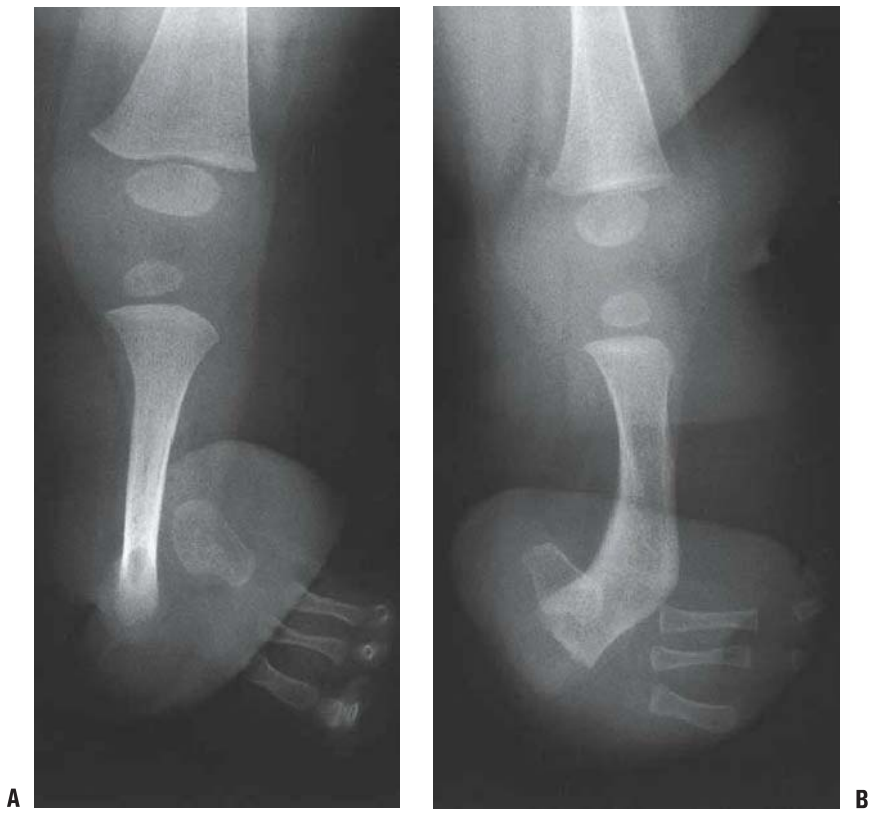


FIGURE 30-8. Type II fibular deficiency. The entire fibula is missing, and there is an anterior tibial bow and missing lateral foot rays.

lateral (postaxial) rays missing, a shortened leg and (often) thigh, a valgus knee, and variable anterior bowing of the tibia with a dimple over the apex. Further examination will demonstrate anteroposterior (AP) instability of the knee (but no instability to varus/valgus stress testing) along with a small patella. In more subtle cases, the only clinical feature may be a limb that is mildly shorter than the contralateral extremity. Although equinovalgus foot deformity is usual, occasionally equinovarus can be seen, especially in these cases (78, 79).

Radiographic Features. Radiographically, the fibula will usually be seen to be shortened in relation to the tibia. This may occur either proximally or distally or both. Often a portion of the fibula is absent in part (Fig. 30-7) or in its entirety (Fig. 30-8). In those cases in which the fibula is of normal or near-normal length, the diagnosis can be difficult during the first year of life. Radiographically, the condylar notch of the femur is shallow and the tibial spines are small.

The femoral shortening may be slight to severe, varying from a few centimeters of shortening to a true PFFD. Amstutz (33) reported femoral deficiency in 15% of those with fibular deficiency, whereas Bohne and Root (80) reported femoral deficiencies in almost two-thirds of their patients. Kalamchi noted that 70% of type I and 50% of type II deficiencies were associated with shortening or deformity of the femur (81).

Radiographs of the knee demonstrate a small hypoplastic lateral femoral condyle (82). The ankle may appear normal in some patients with mild deficiencies, but by skeletal maturity there is usually a valgus deformity. The classic appearance of the ball-and-socket ankle joint in these deficiencies is seen in Figure 30-6C,D. There is disagreement about the origin of this abnormality. Some authors feel that it is congenital (83), whereas others feel that it develops secondary to the tarsal coalitions (84). If it is caused by the tarsal coalition, it is difficult to explain its absence in tarsal coalitions without fibular deficiency. In slightly more severe cases, if the fibula is present, it is short and may not reach the level of the ankle joint. The distal tibial epiphysis shows a triangular appearance.

Although not seen on radiographs at birth, tarsal coalitions are present in most of the feet associated with fibular deficiency. Grogan et al. (85) noted such coalitions in 54% of anatomic specimens, although the abnormality could be seen radiographically in only 15% of the specimens because of the cartilaginous nature of the coalition. Finally, radiographs demonstrate missing lateral rays, which are easily seen clinically without radiographs.

Pathoanatomy. The femoral intercondylar notch narrowing and the tibial spine hypoplasia are indicative of dysplasia of one or both cruciate ligaments. Recently, Manner et al. have correlated the degree of plain radiographic abnormality to the absence of one or both cruciate ligaments as seen by magnetic resonance imaging (MRI) (86). In some cases of fibular deficiency, there is a residual fibrocartilaginous anlage present. Whether or not this anlage causes progressive tibial deformity with growth is unresolved in the literature. Some authors suggest it acts as a growth tether and recommend it be surgically removed primarily (87). Other studies showed no evidence that resection of the anlage made a clinical difference (88).

Natural History. In general, the shortening seen in these patients is progressive according to the rule of proportionality as previously mentioned. With regard to the valgus knee, the deformity usually worsens with growth (36, 80, 89, 90). In some cases, the degree of valgus is more severe than can be explained by the smaller lateral femoral condyle alone, and its recurrence after correction speaks of a more dynamic cause. There is inconclusive evidence to suggest that the anteromedial tibial bow, which is occasionally seen, changes over time. However, clinical experience with untreated older children with fibular deficiency often reveals that the valgus deformity of the ankle progresses over time and can become painful. This is likely a consequence of both the lack of lateral ankle supporting structures and a growth asymmetry at the distal tibial physis. It can also be secondary to a progressive valgus deformity in the mid-diaphysis of the tibia in rare cases where the fibular anlage acts as a lateral growth tether.

Treatment Recommendations. The main problems in the treatment of fibular deficiency are the limb-length discrepancy and the deformity and instability of the foot and ankle. It is very important to realize that the discrepancy will become worse with growth, and it is the ultimate discrepancy at maturity that is important.

Nonsurgical Treatment. Although the majority of patient with fibular deficiency are treated surgically, there are rare cases that are not. If the child has a functional foot without significant valgus deformity, and the degree of shortening would result in an ultimate discrepancy of <2 cm, then no surgery is indicated. Rather, a contralateral shoe lift and serial monitoring during growth for progressive knee or ankle deformity is the preferred treatment.

Surgical Treatment. For the vast majority of patients with fibular deficiency, surgical management is indicated. The great difficulty in treating these children is not deciding which patients would benefit from surgery, but rather which surgical treatment pathway is best for any one patient.

Amputation versus Lengthening. Until the 1960s, amputation for fibular deficiency was recommended only as a last resort (91). In a reaction to the results of these early attempts to save the limbs, several reports emphasized the advantages of amputation for severe cases (36, 92–94). The indications are based primarily on the difference in length and the functionality of the foot. Wood et al. recommended amputation for: a discrepancy of 3 in. or more at the time of decision, or if predicted at maturity; for a nonfunctional foot; for a limb that would have severe cosmetic or functional problems; or for children who cannot endure the psychological trauma of repeated hospitalizations and surgery (94). These recommendations were reaffirmed in a

later publication from the same institution, following up many of the same patients (36).

More recent recommendations begin to stretch the extent to which length can be restored, reflecting improvements in limb lengthening. Westin et al. (36) suggested amputation for any discrepancy that would be >7.5 cm at maturity (36). For Letts and Vincent (21), the number was >10 cm, and for Hootnick et al. (95) it was between 8.7 and 15 cm.

Although modern prosthetics have made amputation a somewhat more acceptable alternative, the improved ability to lengthen limbs has also made limb salvage a more feasible option. The recommendations of Birch et al. are an effort to account for these changes (76). They would recommend amputation for those with a nonfunctional foot, regardless of leg length, unless the upper extremities were nonfunctional. For those with a functional foot, but a leg-length discrepancy of 30% or more, amputation would be recommended. For those with a functional foot and a discrepancy of <10%, epiphysiodesis or lengthening is reasonable. There is little disagreement about these indications today.

It is between those two groups that the controversy regarding treatment lies, and the greater the discrepancy in length, the greater the controversy. According to Birch et al., those patients with a functional foot and a discrepancy between 10% and 30% are candidates for either amputation or lengthening (76). The parents, who are the decision makers, are weighing the hope for their child to retain the limb against what that will entail. Without knowing what a child with an amputation and prosthesis versus a lengthened limb is like, their first response is almost always to lengthen the limb. They most likely have never seen a child or adult with an amputation; they visualize something horrible. At the same time, they cannot really know what a lengthened limb will be like at the end of treatment; they imagine the limb will be normal. Although they may understand that they will need two or three lengthening procedures, they cannot know what the impact will be on their child or their family, what complications they will encounter on the way, or how their child will look or function at the end of the treatment.

As yet, there are but a few preliminary reports of lengthening in fibular deficiencies with predicted discrepancies >10 cm. These preliminary reports, using the Ilizarov methods, deal mainly with the extent of length achieved, often before maturity, but with little information on cosmetic and functional result (96–100).

One way to begin to assess the problem is to look at what amount of length is required. The combined femoral and tibial length for a girl of average height at maturity will be approximately 80 cm (37) (Table 30.2). A 10% discrepancy would be approximately 8 cm, a 20% discrepancy would be 16 cm, and a 30% discrepancy would be 24 cm. To achieve >10 cm of length in a congenital limb deficiency with AP knee instability, ankle instability, foot deformity, and congenitally short soft tissues are a significant undertaking (100–102).

Reports comparing Syme amputation with lengthening are few and incomplete, but begin to give an appreciation of the problems associated with lengthening severe deficiencies (71, 103–105). These reports conclude that lengthening should be reserved for those with more normal feet and less discrepancy in length, although early Syme amputation is the best treatment for the more severe problems. Herring gives a philosophical perspective on the dilemma (106). Birch et al. (107) reviewed a series of adults who were treated with Syme amputation in childhood. These authors conducted physical examination, prosthetic assessment, psychological testing, and physical performance testing and commented that the results of multistaged lengthenings for this condition would have to match these results to be justified. They currently offer lengthening to patients whose limb-length discrepancy is 20% or less.

Bilateral. In patients with bilateral fibular deficiency, the three problems are the foot deformity, the discrepancy in length between the two limbs, and the overall shortening in height because of two short limbs. Without extenuating circumstances (e.g., nonfunctional upper extremities), disarticulation of the foot and prosthetic fitting is the best solution. For those children with nonfunctional upper extremities who will use their feet for many of the activities of daily living (ADL), amputation of the feet is not an option.

In children with bilateral fibular deficiency, there is usually little discrepancy between the two limbs, but rather a discrepancy between their height and what their normal height should be. As they enter into their peer group, this becomes an increasing problem. This problem is most easily solved by the prosthetist. If there is a significant difference between the length of the two limbs that cannot be solved by prosthetic adjustment, lengthening of the short limb becomes an attractive option.

Syme and Boyd Amputation. The amputation described by Syme (108) seems to have been accepted for adults before it was accepted for children, and its use in boys was advocated before its use in girls because it was said that the Syme amputation produced an unsightly bulkiness around the ankle. This resulted in many children receiving a transtibial amputation rather than a Syme amputation. It was subsequently learned, however, that the ankle does not enlarge following amputation in a young child, and the cosmetic appearance is excellent as the child grows.

Thompson et al. were the first to recommend the Syme amputation, rather than transtibial amputation, although only as a last resort (91). Subsequent reports by Kruger and Talbott (93) and Westin et al. (36) not only confirmed the advantages of the Syme amputation in both boys and girls but also advocated its early use for severe deficiencies. Several studies now confirm the value of Syme amputation (90, 93, 106, 109–113).

One of the major advantages of the Syme amputation is the ability to bear weight on the end of the residual limb. This is important both for prosthetic use and for instances in the home when the child will walk short distances without the prosthesis (for instance, going to the bathroom in the middle of the night). It is also relatively technically easy to perform.

In the most complete study to date on the outcome of Syme amputation in children, Herring et al. examined the functional and psychological status of 21 patients with a Syme amputation (Figs 30-9 to 30-14). They noted that family stress was the factor that had the greatest influence on the patients' psychological functioning and that children who had the amputation after several failed attempts at salvage were at considerable risk for emotional disturbance. Green and Cary (114) found that patients were able to function at the average levels for their age group, and the authors did not find that adolescents were less likely to participate in athletics (114). In summary, these studies indicate that Syme amputation may be compatible with the athletic and psychological function of a nonhandicapped child.

A variation of Syme amputation was described by Boyd (115). In the Boyd amputation, the talus is excised and the retained calcaneus with the heel pad is fused to the tibia. The surgery was initially devised to avoid the complication of posterior migration of the heel pad seen in some children with Syme amputation. Advantages of the Boyd amputation are that the heel pad tends to grow with the child, rather than remaining small as in the Syme amputation. In addition, the contour of the retained calcaneus improves prosthetic suspension. The Boyd amputation also adds length. This can be a problem when children who do not have significant shortening of the limb are fitted for various prosthetic feet and may require a shoe lift on the normal side. However, if the residual limb is short enough to fall at the level of the contralateral calf, a Boyd amputation can easily accommodate an energy-storing prosthetic foot, and the added bulk of the residual limb end is easily hidden in the prosthesis.

Eilert and Jayakumar (110) compared the two surgeries and found the migration of the heel pad to be the only complication in the Syme amputation, whereas the Boyd amputation had more perioperative wound problems and migration or improper alignment of the calcaneus. Fulp and Davids (88) compared Syme amputations to a modified Boyd amputation (where the distal tibial epiphysis and physis were removed and the calcaneus was fused to the distal tibial metaphysis). By removing the distal tibial physis and epiphysis, the residual limb was appropriately short, the heel pad was stable, and prosthetic suspension was improved.

SYME AMPUTATION (FIGS. 30-9 TO 30-14). The Syme amputation in congenital deficiencies in children has two important differences when compared to adults. First, in children with severe congenital deficiency of the lower extremity, the foot is often in severe equinus, with the heel pad proximal to the end of the tibia. This may result in difficulty in bringing the heel pad down over the end of the tibia, even after sectioning of the Achilles tendon. Second, no bony alteration of the distal tibia is necessary. The malleoli are not a problem with prosthetic fitting because they do not attain the usual medial and lateral dimensions of the adult (90). In fact, a slight prominence is necessary for suspension of the prosthesis.

The most often cited benefit of this amputation is the end-bearing ability of the stump, which permits walking without a prosthesis and better prosthetic use. This end-bearing quality is dependent on the preservation of the unique structural anatomy of the heel pad by careful subperiosteal dissection of the calcaneus. One of the most obvious benefits of a Syme amputation (or any disarticulation) in childhood is the elimination of bony overgrowth, with the necessity for revision that accompanies through-bone amputation in the growing child. Although there are many reports of the long-term results in patients undergoing the Syme amputation, most of these have been performed for other indications. (116, 117).

BOYD AMPUTATION WITH OSTEOTOMY OF THE TIBIA FOR FIBULAR DEFICIENCY (FIGS. 30-15 TO 30-18). This amputation, first described by Boyd in 1939, is best indicated in the limb-deficient child. The amputation is similar to the Syme amputation except that it preserves the calcaneus with the attached heel flap and fuses it to the distal tibia. In the congenitally deformed foot found in congenital lower extremity deficiencies, the arthrodesis might favorably affect the fixation and the growth of the frequently occurring small heel pad, leaving the heel pad intact on the calcaneus. Its disadvantage is that in these same patients, the calcaneus and the distal tibia are largely cartilage, making arthrodesis difficult to achieve. If arthrodesis is not achieved, the calcaneus will migrate from beneath the fibula, requiring revision or conversion to a Syme amputation, which is not required when the heel pad alone migrates. The procedure, although most commonly used in the treatment of fibular deficiencies, has also been used in the treatment of tibial deficiencies by fusing the calcaneus to the fibula.

Correction of Tibial Bow. The anterior bow in the diaphysis of the tibia varies from nonexistent to severe. Severe bowing is usually seen in the more severe deficiencies with complete absence of the fibula. Westin et al. reported this to be of little consequence (36). However, observations in the authors' center have shown this to be an occasional prosthetic problem, requiring osteotomy during the first decade.

With the tibial bow, the foot is displaced posterior to the weight-bearing axis that passes through the knee. If the foot is placed at the distal end of the tibia (which the parents want for cosmetic reasons), the ground reaction force places a large moment through the toe-break area, leading to premature failure of the foot component and skin problems caused by abnormal pressure. The problem is then blamed on the foot component or the prosthetist.

A reasonable recommendation would be to correct any significant bow at the time of Boyd amputation. A small anterior incision, removal of an anterior-based wedge of the tibia, and fixation with a temporary Steinmann pin placed up through the heel pad and calcaneus, and crossed Steinmann pins placed at the level of the osteotomy solves the problem and does not result in any delay in prosthetic fitting

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Syme Amputation (Figs. 30-9 to 30-14)

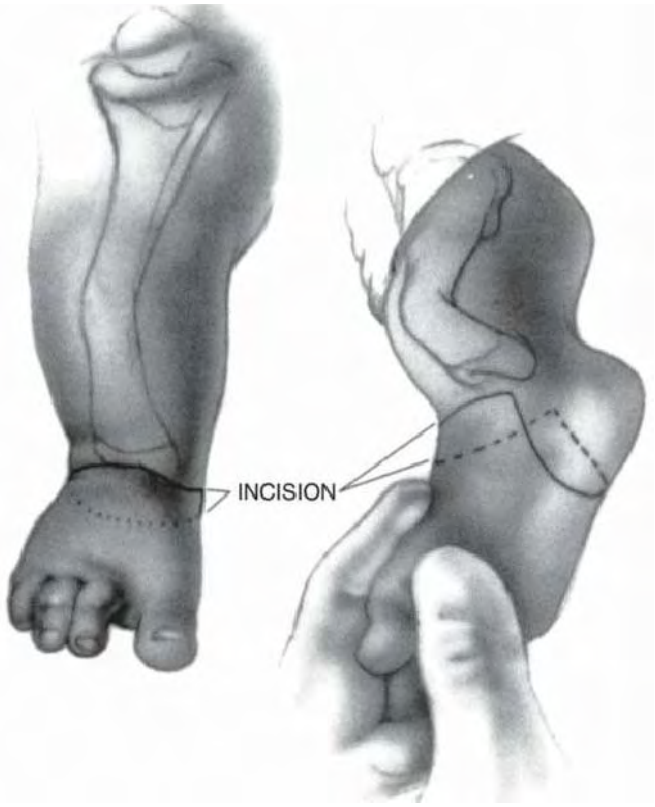

FIGURE 30-9. Syme Amputation. The dorsal part of the incision begins at the tip of the lateral malleolus, crosses over the dorsum of the foot near the dorsiflexion crease of the ankle, and ends about 1 cm below the tip of the medial malleolus. In children with complete absence of the fibula, this landmark can be estimated by palpation of the anatomy, as well as visually. The volar part of the incision connects the ends of the dorsal incision crossing the plantar aspect of the foot at the distal end of the heel pad. In young children who have not been walking, the heel pad is often difficult to identify, and care should be taken to bring the incision far enough distally to retain sufficient tissue.

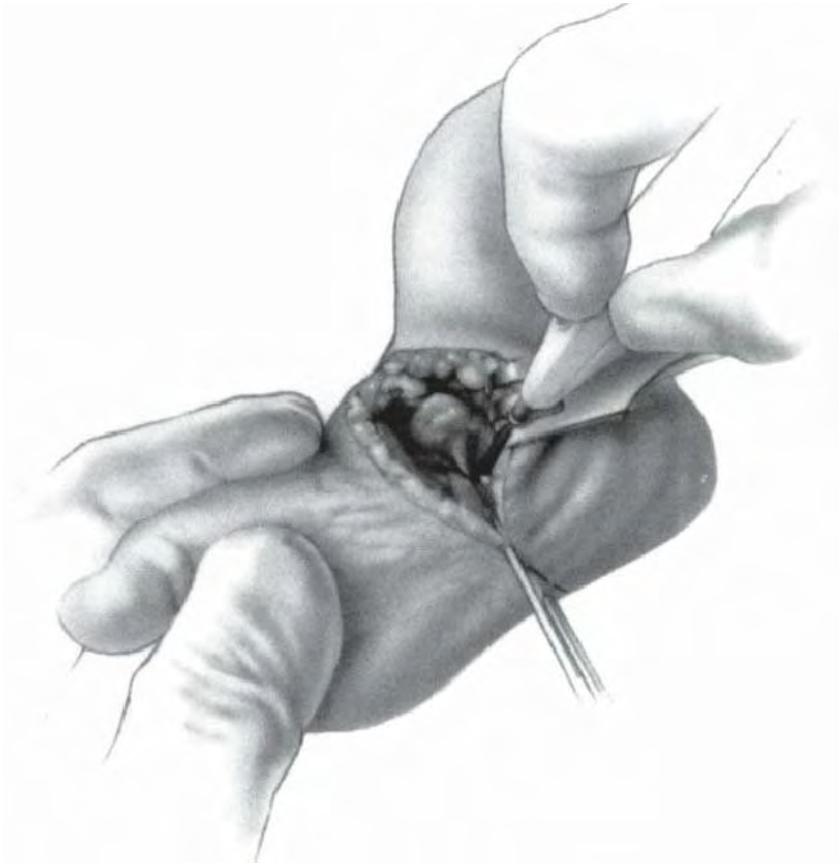

FIGURE 30-10. The next step is to complete the plantar incision. Because nothing distal to this point will be saved, this incision can be carried directly down to the bone, identifying and cauterizing the bleeding vessels later when the tourniquet is released. The tendons and nerves can be pulled distally and sectioned so that they retract proximally and the vessel can be ligated or cauterized. Completing this part of the incision simplifies the most difficult and important part of the operation, which is to divide the medial and lateral ligament structures without injuring the posterior tibial vessel and its branches that supply the heel pad.

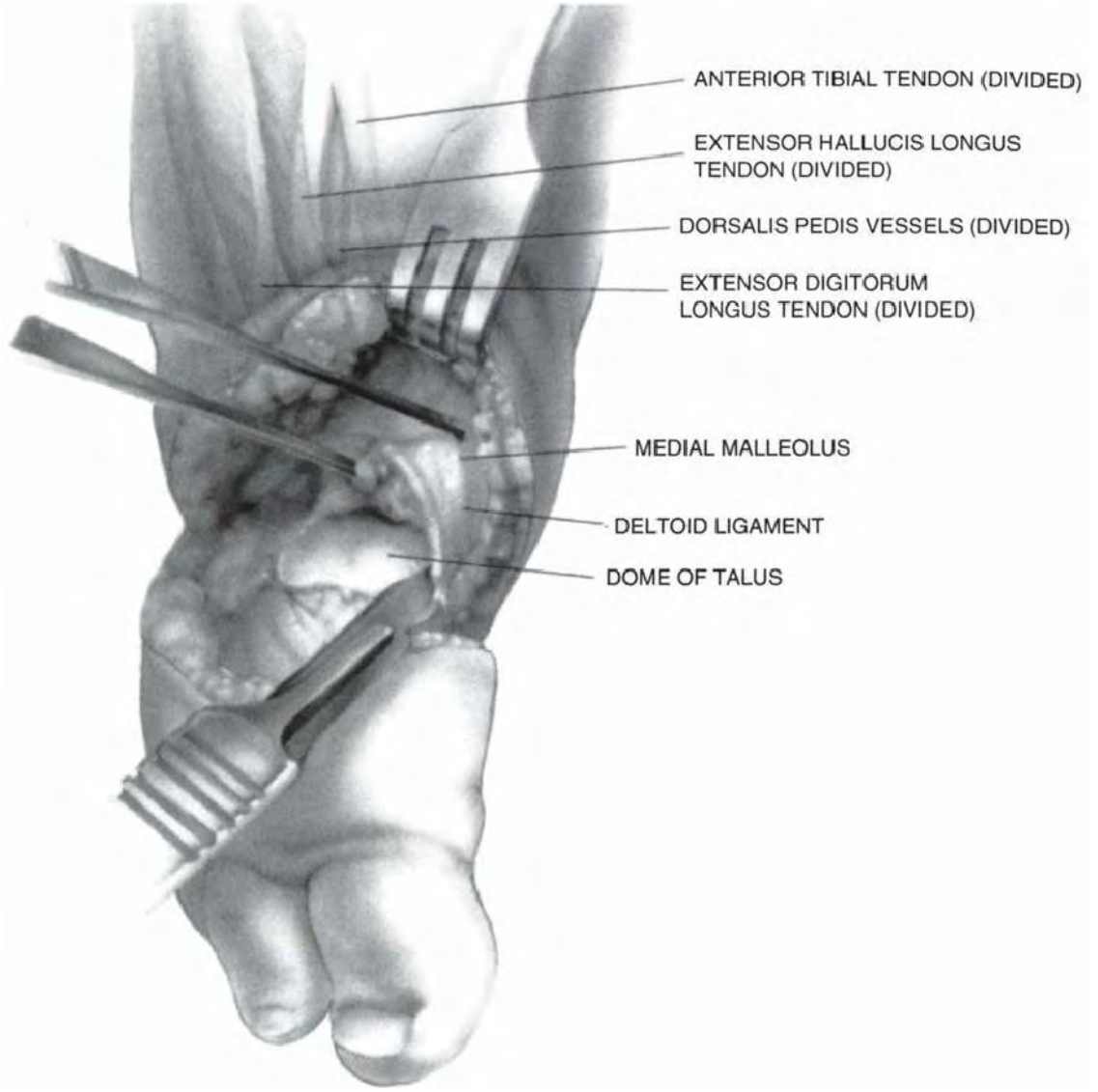

FIGURE 30-11. Next, the dorsal incision is deepened through the subcutaneous tissue. The dorsalis pedis vessels are identified and cauterized, and all the tendons and nerves are pulled distally, sectioned, and allowed to retract proximally. This exposes the anterior ankle joint, which is now cut open completely. Working carefully between the talus and the medial malleolus, the deltoid ligament is cut, freeing the medial aspect of the ankle joint. Care is necessary here to avoid damage to the posterior tibial artery and vein. Working on the lateral side of the ankle, the surgeon cuts the tibiofibular ligaments. The only remaining portion of the ankle capsule remaining is posterior.

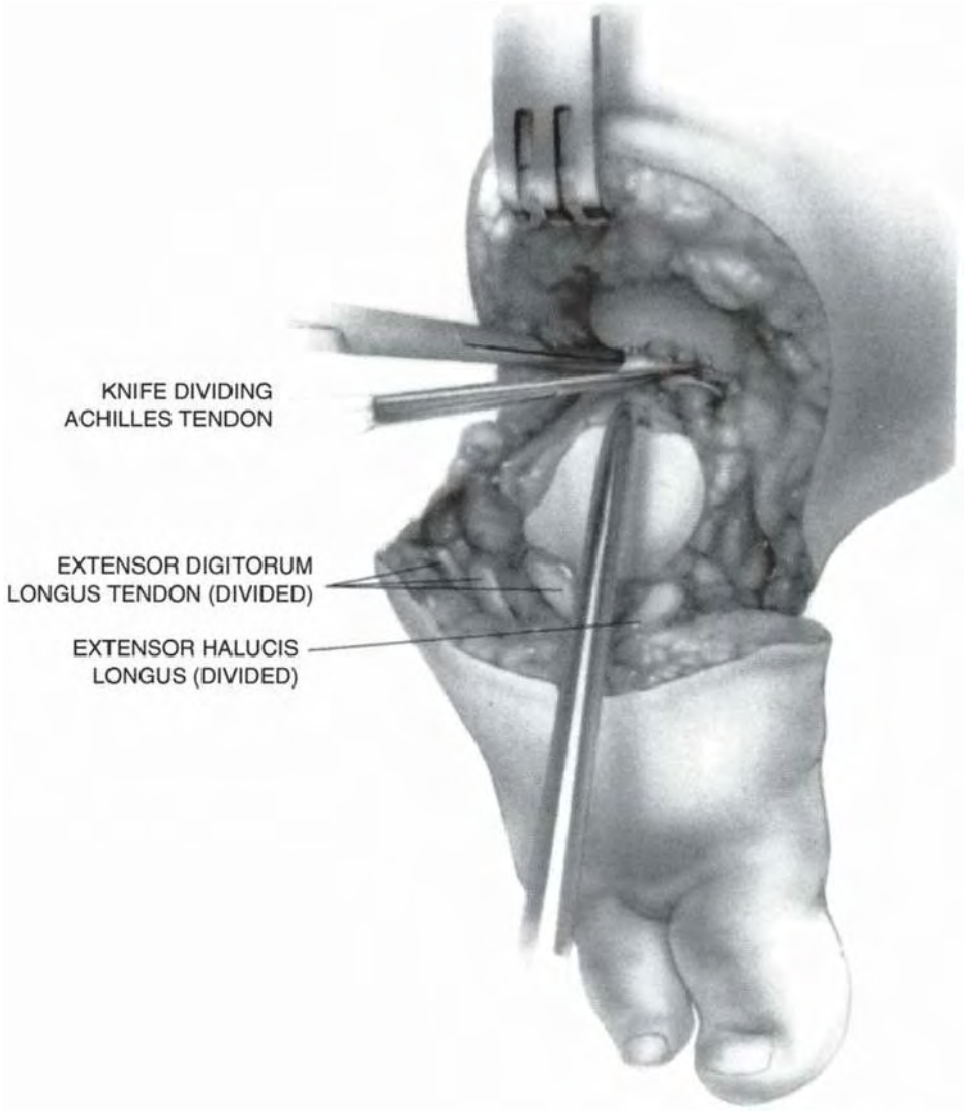

FIGURE 30-12. With the foot severely plantarflexed and a small bone hook placed in the posterior aspect of the talus, the dissection of the posterior ankle capsule is completed, freeing the talus. In the severe congenital foot deformity, this is not easy, and care must be taken to avoid cutting through the cartilaginous portion of the posterior talus. When this part of the release is completed, the talus should come forward, exposing the calcaneus.

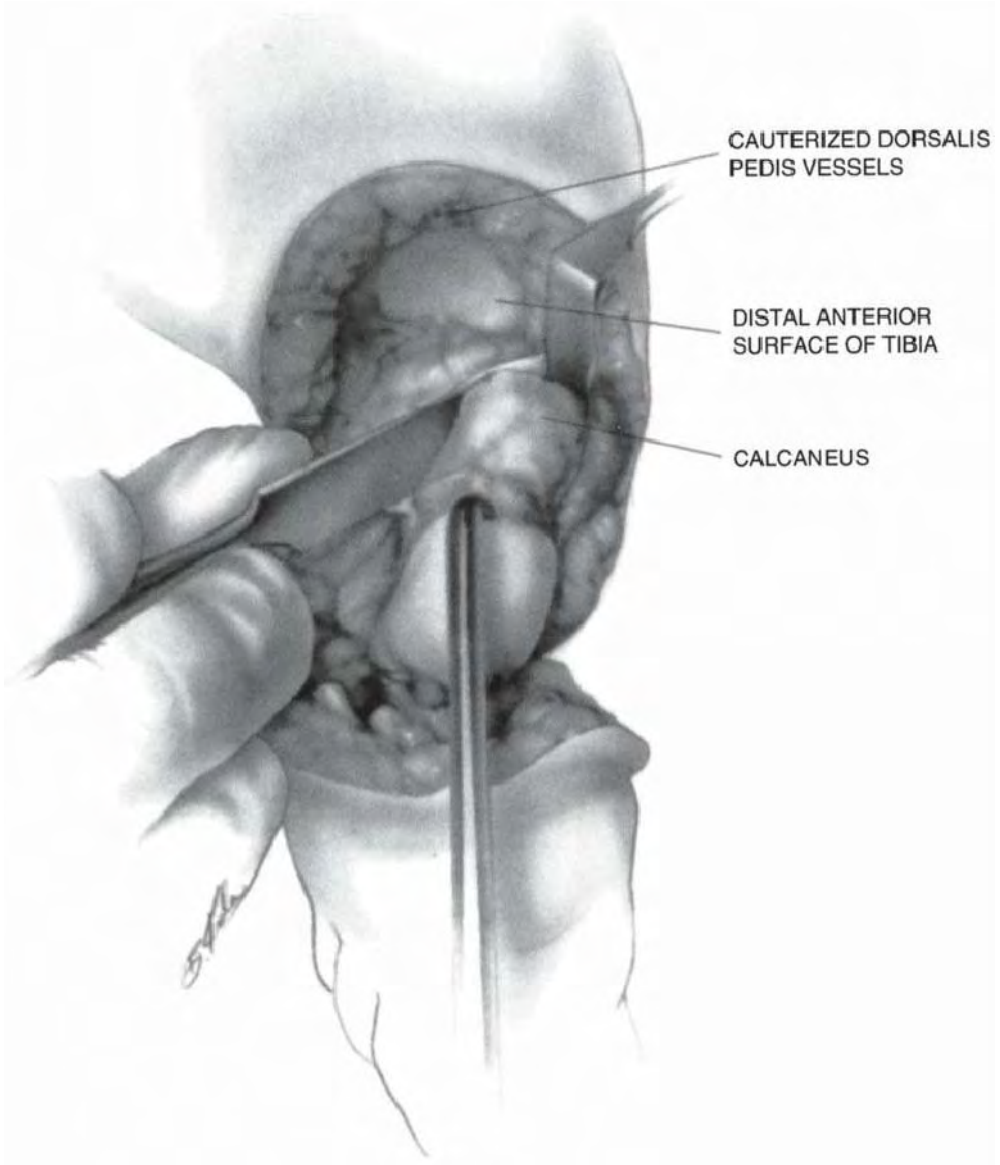

FIGURE 30-13. The dissection is completed with subperiosteal removal of the calcaneus. Using both a scalpel and periosteal elevator, the dissection is started posteriorly from within the ankle joint. Here again, the surgeon must be careful not to separate the calcaneal apophysis from the calcaneus. As soon as the Achilles tendon is identified, it should be exposed and a portion of it removed. This eliminates the tendency of the gastrocnemius muscle to pull the heel pad off the tibia, a common complication. After dividing the Achilles tendon, the dissection of the calcaneus proceeds around to the plantar surface. In some congenital anomalies, however, in which the heel is behind the tibia, it will be much easier to start dissecting the calcaneus from the distal to the proximal side.

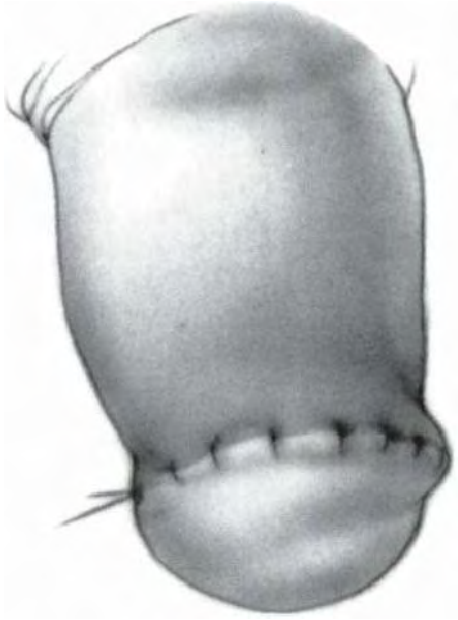

FIGURE 30-14. It is not necessary to remove the cartilage from the distal tibia or to cut any bone from the distal tibia. Closure is accomplished by suturing the deep layer of the heel pad to the deep fascia and periosteal tissue of the proximal part of the dorsal incision. This will anchor the heel pad. Fixation pins are not necessary and are largely ineffective because posterior migration of the heel pad occurs late as a result of the imbalance of forces created by the failure to section completely all tendons pulling on it. It is wise to deflate the tourniquet before closure in order to check the circulation to the flaps and to control the bleeding. Drains do not appear to be necessary.

(Fig. 30-19A–E). When correction of the bow is necessary in older children who are already in their prosthesis, several fixation options, including crossed Steinman pins, plates, or intramedullary devices are available.

Correction of Genu Valgum. Development of progressive genu valgum has long been known as a complication of fibular deficiency. It is one of the major problems seen in the gait of children with this problem. At first, it is merely cosmetic and can be accommodated with prosthetic alterations. However, if it becomes more severe, it will increase the forces on the lateral compartment and make good alignment impossible.

Westin et al. noted that the tibia often developed an anterior flexion along with the valgus, and attributed the problem to an abnormality in growth in the lateral and posterior portions of the proximal tibial physis (36). This problem is different from anterior bow in the diaphysis of the tibia.

Most recently, Boakes et al. (82) documented a decrease in the height of the lateral femoral condyle that was not present prior to walking. There was a suggested relation between the extent of anteromedial bowing of the tibia and the subsequent decrease in height of the lateral femoral condyle. They suggested that tibial osteotomy might prevent the changes in the lateral femoral condyle and correct the anteromedial bowing. If the deformity was present in the lateral femoral condyle, they suggested temporary stapling of the medial femoral condyle, since osteotomy has a very high recurrence rate unless performed near the end of growth. The authors' experience indicates that it is not as simple as this and that the recurring nature of the valgus following good correction of alignment suggests other causes of this problem.

Ankle Reconstruction. Any attempt to save the limb of a child with significant fibular deficiency will require efforts to realign and stabilize the ankle. There is renewed interest in this subject with attempts to lengthen the leg.

The Gruca procedure is designed to provide lateral stability to the foot in the absence of the fibula. Serafin gives the first report of the technique in the English literature and recounts the various attempts at bone grafting and other procedures that were described before Gruca developed his technique (118).

In the Gruca procedure the tibia is split longitudinally. The medial segment is displaced proximally with the talus, leaving the lateral fragment as a lateral buttress. Thomas and Williams describe the early results in nine patients treated with this procedure. The follow-up is short and the evaluation of function incomplete (119). More recently, a newer procedure was described to provide lateral stability that involves transplantation of a tricortical iliac crest graft with apophysis and gluteal fascia to the lateral distal tibia (120). Neither surgery has been widely used and would seem to have little to recommend it.

Arthrodesis of the talus to the distal tibia is a logical plan in conjunction with leg lengthening, but there are no reports on its outcome. It is likely that this would also require release of all of the tendons crossing the ankle joint to prevent foot deformity. Drift of the foot through the physis or the fusion itself with lengthening and over time seems a possibility. There are also case reports of tibial lengthening with an extra-articular screw from the calcaneus to the tibia to prevent progressive equinus (121). Neither technique can be recommended at this time.

The ball-and-socket ankle joint, seen in the Kalamchi type IA deficiencies, usually require no treatment. The authors have, however, seen several children with increasing valgus during adolescence or following leg lengthening who become symptomatic with normal athletic activity. They have been successfully treated by a distal tibia varus angular osteotomy.

Prosthetic Management. Prosthetic management of the fibular-deficient limb is different than management of a Syme amputation in an adult after trauma. In the child, the prosthesis is designed to accommodate growth and to help stabilize knee laxity and hyperextension through socket design and alignment. Emphasis is placed on socket alignment and minimizing rotational forces acting on the knee.

The socket fitting for a Syme or Boyd amputation may be designed to bear all of the weight on the end of the residual limb, as intended, on the patellar tendon and flare of the proximal tibial condyles, as in a transtibial amputation,

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Boyd Amputation with Osteotomy of the Tibia for Fibular Deficiency (Figs. 30-15 to 30-18)

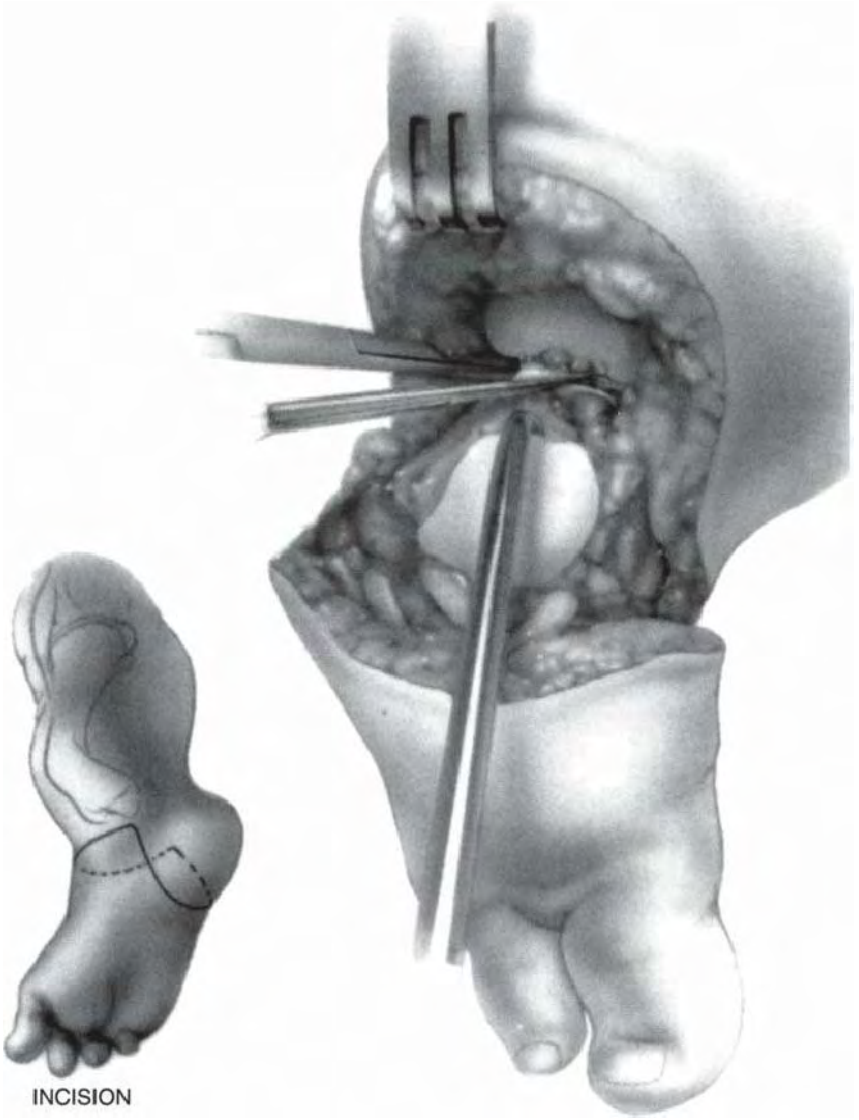


FIGURE 30-15. Boyd Amputation with Osteotomy of the Tibia for Fibular Deficiency. The incision and the initial exposure for the Boyd amputation is the same as for the Syme amputation through the release of the posterior ankle capsule. At this point, the foot is amputated through the midtarsal joints (calcaneocuboid and talonavicular). Because the forefoot provides a good handle by which to control the hindfoot, it is easiest to dissect most of the talus free and section the Achilles tendon, if possible, before removing the forefoot.

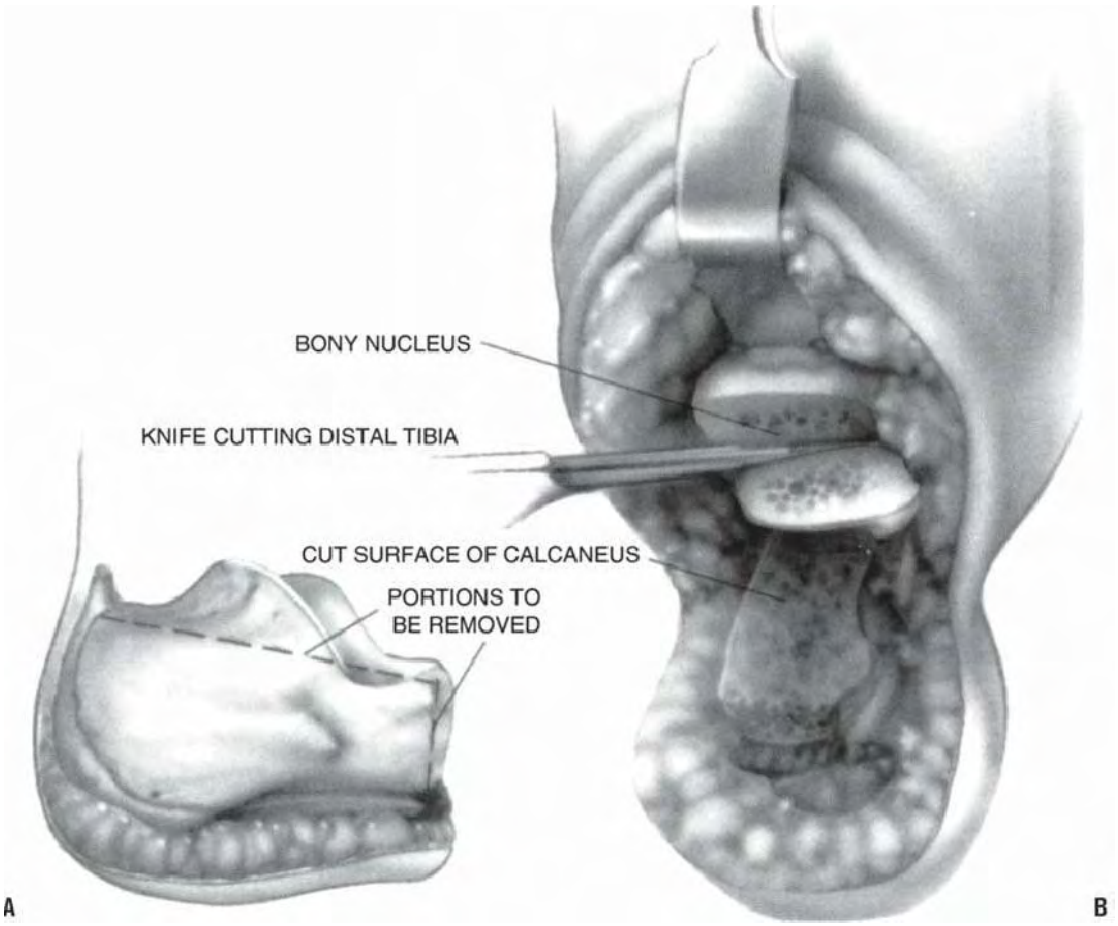


FIGURE 30-16. The talus is completely removed by cutting through the talocalcaneal ligaments. A saw (or often only a knife) is used to fashion a portion of the calcaneus to fit under the articular surface of the tibia. First, a portion of the anterior calcaneus is removed. Next, the articular surface of the calcaneus is removed to expose cancellous bone **(A)**. Finally, enough of the distal tibia is resected to expose the bony nucleus of the distal epiphysis **(B)**. A rongeur is used to shape the calcaneus so that good bony contact is achieved between the calcaneus and the distal tibia. This is stabilized by a smooth Steinmann pin that passes through the heel pad and the calcaneus and into the tibia. The calcaneus must be pulled forward before fixing it to the tibia to avoid a posterior prominence that would interfere with prosthetic fitting and wearing.

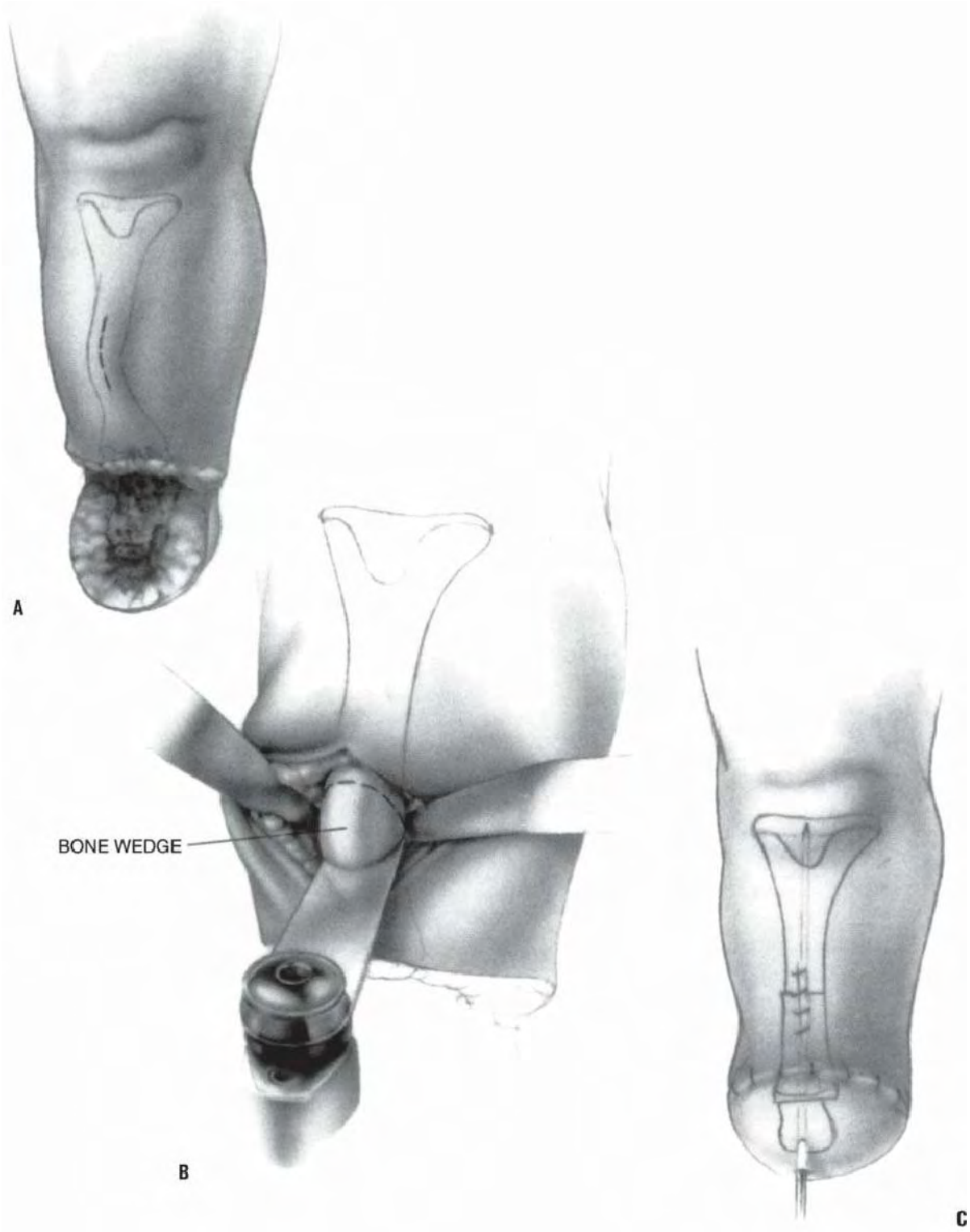


FIGURE 30-17. In many cases of fibular deficiency, the tibia is bowed anteriorly. Although this does not cause a problem with prosthetic fitting during the child's younger years, it may worsen with growth, making both prosthetic fitting and alignment of the foot difficult later. If it appears that this deformity will be significant, it is best to correct it at the same time as the amputation (Syme or Boyd) because it does not delay fitting significantly and at this age is preferable to a tibial osteotomy in later childhood. A straight incision is made directly over the subcutaneous border of the tibia at the apex of the bow and carried directly down through the periosteum **(A)**. A small area of the tibia is exposed subperiosteally, leaving the posterior periosteum intact. A saw or rongeur is used to remove a small wedge of bone in the appropriate plane to correct the deformity. Usually this is not a straight anterior bow **(B)**. A Steinmann pin is passed through the heel pad, through the calcaneus, into the distal fragment of the tibia, and then into the proximal fragment of the tibia. The wounds are then closed **(C)**.

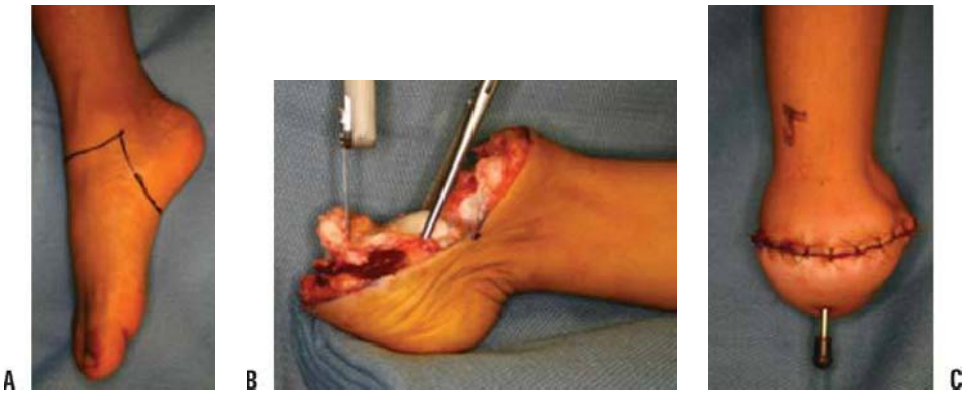
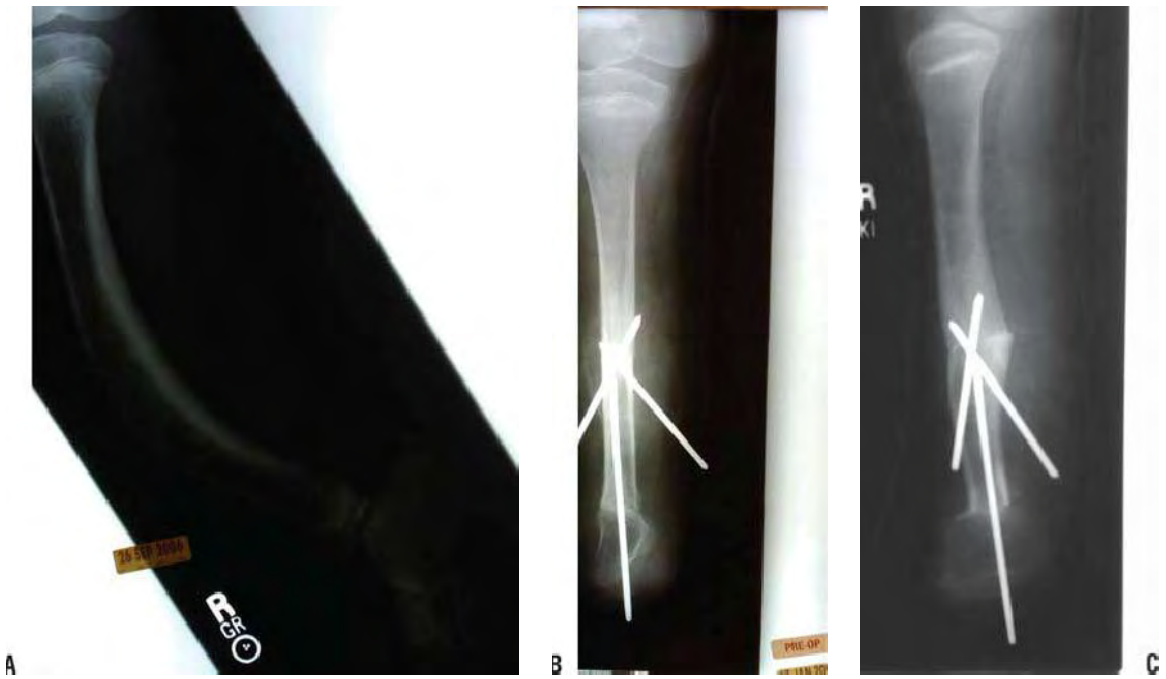


FIGURE 30-18. **A–C**: Pre-, intra-, and postoperative clinical photos of a patient with fibular deficiency undergoing Boyd amputation. Note the threaded Steinman pin protruding through the heel pad, which is used to fix the calcaneus to the distal tibia to promote fusion.



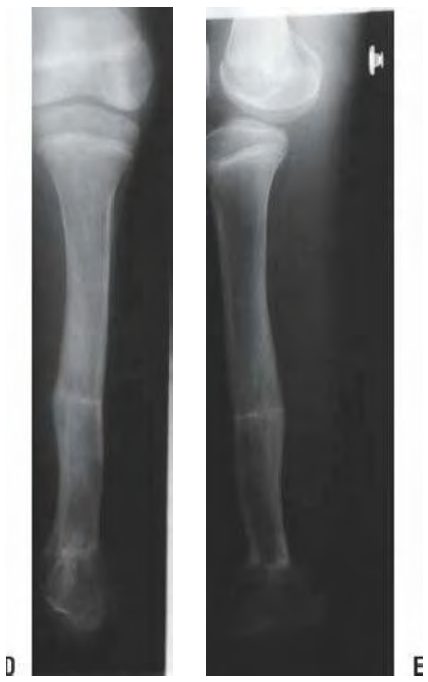


FIGURE 30-19. **A**: Anteroposterior radiograph of a type II fibular deficiency (Achterman and Kalamchi), in which the entire fibula is missing. Note the missing lateral rays of the foot and the severe angulation of the tibia. **B,C**: The limb, 6 weeks after Boyd amputation and an anterior closing-wedge osteotomy of the tibia. Placing the pin through the anterior cortex of the proximal fragment provides rigid fixation, which is not obtained if the pin is simply passed up the medullary canal. The pin was removed in the office at the time of cast removal. **D,E**: The clinical appearance of the same deficiency at the time of surgery in another patient. Note the short tibial segment, the valgus knee and foot, and the dimple over the tibia.

or both. As the child grows, it is a good idea to begin shifting some of the weight bearing to the proximal structures to prepare the child for the time when full weight bearing on the end may not be possible. Failure to shift weight bearing proximally with age usually leads to fitting difficulties resulting from tolerance issues. This discomfort probably arises from the small distal weight-bearing surface seen in many of the congenitally deformed limbs. This may be partially alleviated with a Boyd amputation, which is generally broader than the Syme amputation. This is an especially important consideration in the bilateral amputee, in whom disproportionate weight shifting to the sound side is not possible for comfort.

As with most amputations done at a young age, the condyles will be small at the time of amputation, will not grow to normal size, and therefore, do not require trimming as in the adult. The proximal brim of the socket is designed with supracondylar (SC) medial and lateral trim lines, in an effort to control any knee valgus instability and/or patellar instability. The type of suspension will depend on the size of the distal end of the residual limb. If it is very large, an obturator or window may be necessary. With further growth, the distal end may not be sufficient for suspension, and a different design will be necessary. These are discussed later.

To best utilize the current prosthetic feet in children who are older and large enough to take advantage of them, it is necessary that at least 4 cm of space be available at the distal end. If the prosthetist is to offer the latest in energy-storing feet, greater residual limb-length differences will be required. This need can be anticipated, and an arrest of the distal or proximal tibial and fibular physes performed at the appropriate time. This length differential is usually not a problem in children with congenital limb deficiency, because the deficient limb will usually be shorter than the other limb. However, it can be an issue in children with acquired deficiency treated by Syme or, more often, Boyd amputations. Although the longer lever arm of the Syme amputee tends to compensate for the lack of more elaborate prosthesis components, when fit is possible, they can be an advantage.

Authors Preferred Recommendations. For patients with a functional foot with at least 3 rays and a predicted discrepancy of <20 cm, the authors suggest retaining the foot with the goal of limb-length equalization. If the discrepancy is projected to be 5 cm or less, a contralateral pan genu epiphysiodesis at the appropriate time is the treatment of choice. For greater discrepancies, limb lengthening is added to the treatment plan. Lengthening is done with a monolateral external fixator unless there is secondary angular or rotational deformity that is being corrected at the same time. In those cases, a circular fixator is preferred. A Boyd amputation is suggested for patients with >17 cm of predicted discrepancy or in patients with a nonfunctional foot. In rare instances where the calcaneus is significantly hypoplastic, or if it is proximally displaced in the posterior calf, a Syme amputation is performed.

For patients with planned lengthening and distal femoral valgus, the authors prefer to correct the valgus simultaneously with femoral lengthening. If no femoral lengthening is planned and the deformity is >10 degrees in a patient with at least 2 years of distal femoral growth remaining, a medial distal femoral reversible hemiepiphysiodesis, with removal of implants when the angulation has resolved. A distal femoral angular osteotomy is reserved for the skeletally mature adolescent with prosthetic fitting difficulties and genu valgum.

Tibial anteromedial bowing is corrected at the time of amputation if it will interfere with prosthetic fitting. In the authors' experience, this is generally true if the angulation is >30 to 40 degrees. In the young child who is undergoing amputation at the same time, the authors fix the osteotomy by running the threaded Steinman pin that crosses the calcaneus and tibia more proximally to fix the osteotomy and engage the proximal anterior tibial cortex. For the older child, we prefer crossed Steinman pin fixation Finally, fibular anlage resection is reserved for the rare patient who has recurrent anteromedial tibial bowing after osteotomy or progressive bowing over time.

Pearls/Pitfalls. The authors have treated several patients who demonstrated a recurrent or progressive anteromedial tibial bow after correction with osteotomy. In these cases, surgical exploration with resection of the fibular anlage has proved a successful treatment.

When initially evaluating the patient with fibular longitudinal deficiency, it is important to carefully examine the contralateral extremity. It can be easy to miss a case of bilateral deficiency when the deficiency on one side is mild. Make sure not to focus on the more abnormal limb and to examine the entire patient.

Complications

Amputation. One of the major complications of the Syme amputation is migration of the heel pad off the end of the residual limb (Fig. 30-20). This is particularly true in congenital limb deficiencies, in which the heel may be on the back of the tibia, making repositioning of the heel pad on the end of the limb difficult or impossible. Heel pad migration can be addressed with prosthetic modifications, but the consequence is that it is no longer an end-bearing amputation. Most other problems in patients with Syme amputation are caused by other effects of the underlying disorder (90, 91, 109, 111).

With regard to the Boyd amputation, the main complication is the migration of the calcaneus if successful arthrodesis is not achieved. Arthrodesis is difficult because both the distal tibial epiphysis and the calcaneus are largely cartilaginous at the time of amputation. This can be addressed by removing the distal tibial epiphysis and fusing the calcaneus to the distal tibial metaphysis, as mentioned previously. However, if pseudarthrosis with calcaneal migration occurs, this can require an additional surgery, which is often conversion to a Syme amputation.

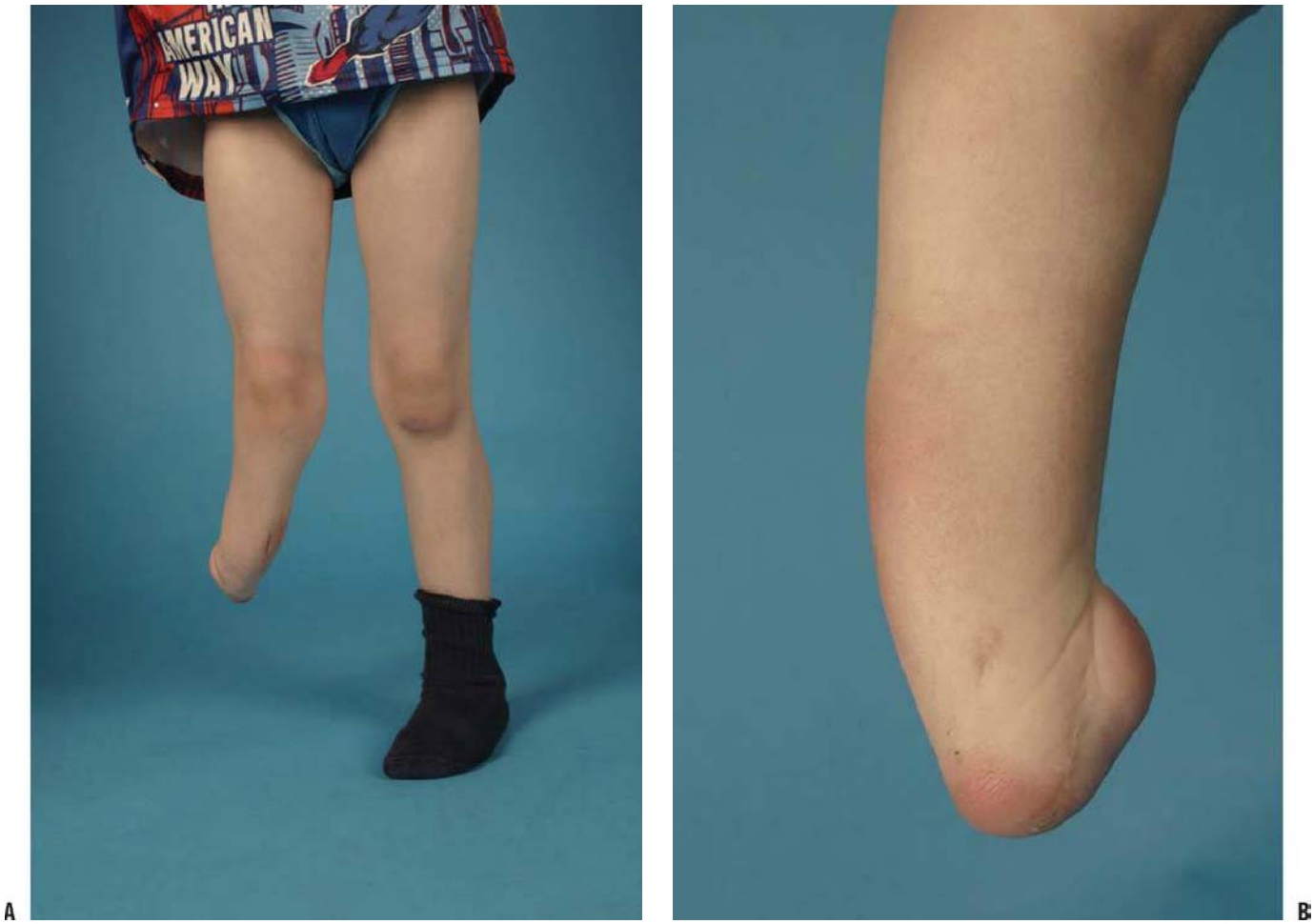


FIGURE 30-20. **A,B**: AP and lateral clinical photos of a patient with fibular absence who had a Syme amputation with heel pad migration. The heel pad has migrated posteriorly, and the patient has a painful discolored area over the distal tibia from weight bearing.

Lengthening. One complication specific to these patients is subluxation of the knee or ankle during lengthening. At the knee, posterior subluxation of the tibia and development of a progressive knee-flexion contracture can be seen with femoral lengthening. This happens because there is a high incidence of cruciate ligament dysplasia in these patients. Prevention of this problem before it occurs is best, which can be accomplished by using a hinged knee orthosis that can be locked in extension, performing an extra-articular anterior cruciate ligament reconstruction, and/or by spanning the knee joint with the external fixator with a hinge placed at the knee joint during lengthening.

Progressive subluxation can also occur at the ankle joint with lengthening of the tibia. These patients often have abnormal lateral supporting structures, including the lateral malleolus and lateral stabilizing ankle ligaments. Inclusion of the foot in the fixator frame is one treatment which has some success in preventing the complication (122). If the complication develops, subsequent ankle soft-tissue stabilization, distal tibia osteotomy, or fusion may be necessary (123).

Tibial Deficiency

Definition and Classification. The most common classification system of tibial deficiencies is the Jones classification, which is a modification of the earlier Kalamchi and Dawe classification (99). The Jones classification is based on the radiographic appearance of the limb within the first year of life. In type 1 tibial deficiencies, ossification of the tibia is absent. In type Ia, there is hypoplasia of the femoral epiphysis, implying a complete lack of any tibial anlage articulating with the distal femur (Fig. 30-21). In these children, the extensor mechanism is usually severely deficient or absent. In rare cases, bifurcation of the distal femur can be an associated finding in these patients. In type 1b tibial deficiencies, the femoral condyles are more developed, implying there is a cartilaginous tibial anlage present that articulates with the distal femur (Fig. 30-22). This anlage eventually ossifies with further growth usually between 2 and 4 years of age. These patients often have a functioning extensor mechanism.

Jones type 2 tibial deficiency has a proximal tibia with absent ossification of the distal tibia. This group is the same

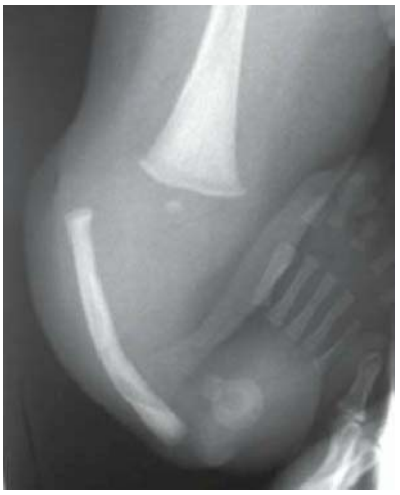


FIGURE 30-21. AP radiograph of the lower extremity in a patient with type Ia tibial longitudinal deficiency. Note the severely delayed distal femoral epiphyseal ossification and absence of tibial ossification.

as type II in the original Kalamchi classification. Jones type 3 tibial deficiency is extremely rare and consists of absence of the proximal tibia with presence of the distal tibia. Finally, in type 4 deficiency, there is diastasis of the distal tibiofibular joint, where the talus is wedged between the distal tibia and fibula and can articulate with the fibula alone or the fibula and tibia.

The presence or absence of intact extensor mechanism function is the primary initial decision point in the treatment of these children. This usually coincides with the presence or absence of a proximal tibia (cartilaginous or bony). This usually (but not always) depends on the presence of a proximal tibial segment. A good radiographic clue is that in patients who have a proximal portion of the tibia, the distal femoral condyle is wider and the ossification of the epiphysis is better than if it is not present.

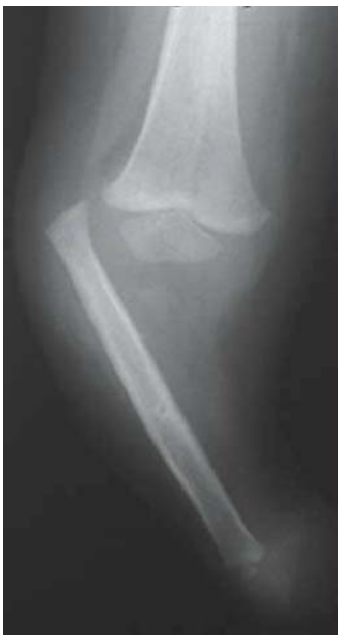


FIGURE 30-22. AP radiograph of the lower extremity in a patient with type 1b tibial longitudinal deficiency. Note the increased ossification of the distal femoral epiphysis as compared to Figure 30-21.

An unusual type of tibial deficiency is that seen in association with fibular dimelia. Kumar and Kruger summarized the sporadic reports until 1993 and presented the findings, associated anomalies, and treatments in six patients (102). In this deficiency, the tibia is absent and there is a duplication of the fibula. These patients have a high incidence of other anomalies, including visceral anomalies. The authors recommended knee disarticulation if the femur is of normal length and fusion of the fibula to create a sufficient lever arm if there is associated PFFD.

Epidemiology and Etiology. Tibial deficiency is much less common than fibular deficiency, with an incidence reported to be 1 in 1,000,000 live births (126). Also in contrast to fibular deficiency, tibial deficiency can be genetically inherited. Although most commonly this is seen as with autosomal dominant inheritance, cases of autosomal recessive inheritance have been reported by several authors (127–129). The majority of patients with tibial deficiency will have associated anomalies of the musculoskeletal and other organ systems, with an incidence of approximately 75% (124, 130, 131). Several well-described syndromes include tibial deficiency among the list of findings (127, 132–134), and genetic counseling is recommended.

Clinical Features. The typical appearance of an infant with tibial deficiency is a markedly shortened tibia with a rigid equinovarus-supinated foot pointing toward the perineum (Fig. 30-23). Preaxial polydactyly is the classic appearance of the forefoot, although the absence of the preaxial rays or a split foot deformity can also be seen. The fibula is prominent at the proximal lateral aspect of the knee, and the knee is most often unstable to AP and varus/valgus stress testing. The examining physician should actively look for other musculoskeletal anomalies, such as preaxial upper limb polydactyly, hip dislocation, or scoliosis.

Radiographic Features. The Jones classification encompasses many of the radiographic features seen in these patients with respect to the lower leg segment. Other findings include proximal dislocation of the fibula into the distal lateral thigh segment, which gives its common appearance on physical exam. As mentioned previously, the distal femoral epiphysis can exhibit delayed or absent ossification in Jones type Ia tibial deficiency.

Other radiographic features are those of commonly associated anomalies, such as hand or foot anomalies (preaxial polydactyly, missing digits, or split hand or foot anomaly), hip anomalies (DDH, PFFD, coxa valga), or spine anomalies (congenital scoliosis).

FIGURE 30-23. Clinical photograph of the lower extremity of a patient with tibial deficiency. Note the equinovarus foot position and shortening of the lower leg as compared to the thigh segment.

Other Imaging Studies. Rarely, there is difficulty determining the clinical presence of an unossified proximal tibia in the infant in Jones 1b tibial deficiency. Some authors have recommended obtaining an ultrasound (135) or MRI to detect its presence. MRI can be useful in this situation because it can identify the rare patient with active knee extension and no tibial remnant, who is treated differently than the patient with active knee extension and a tibial remnant.

Pathoanatomy. Pathoanatomy has been determined from specimens at the time of knee disarticulation. Common findings include an anomalous fibrotic band attached via interosseus membrane to the fibula, the presence of unidentified muscle attached to the tip of the lateral malleolus, and the duplication of anterior compartment muscles. Abnormal cruciate ligament, collateral ligament, and menisci are often seen. The patella is always abnormal and often absent, with the absence of the patellar ligament. Popliteal artery division has been described as being more distal than normal, with bifurcation at the midfibular level (129, 136). Lack of a normal-appearing tibial nerve, with a lateral nerve coursing down the lower leg and dividing in the sole of the foot, have also been found (137).

Natural History. There are no reports on the natural history of tibial deficiency. In bilateral cases of type Ia deficiency, ambulation is not observed. The authors have observed several patients with bilateral types 2 and 3 tibial hemimelia who presented for treatment between the ages of 5 and 8 years. These children were ambulatory but had complaints of pain due to foot deformity or proximal fibular prominence.

Treatment Recommendations

Nonsurgical Treatment. Nonsurgical treatment is only indicated in the patient with bilateral tibial deficiency with active knee extension and acceptable foot position. Any other patient with tibial deficiency stands to gain much by the various types of surgical treatment.

Surgical Treatment. The treatment of Jones type Ia tibial deficiency without active knee extension is knee disarticulation and prosthetic fitting. However, there are several reports in the literature of attempts to centralize the fibula between the femoral condyles, which are discussed below. Originally described by 1905 by Myers (128) and subsequently by Sulamaa and Ryoppy in 1963 (138), the Brown procedure, as it is commonly known in the United States, is the centralization of the fibula under the femur (139, 140). It was Brown's recommendation that fibular centralization be done only with active knee extension, which is rarely seen in the case of a completely absent tibia. Most reports on the Brown procedure, however, include significant numbers of patients who did not have well-documented active knee extension before surgery. Some reports also erroneously include patients who actually had transfer of the fibula to a proximal tibial remnant rather than complete transfer of the fibula under the femoral condyles. The surgery has now been evaluated in several clinical trials (132, 133, 141–144). Most of those reporting on the procedure recommend against it, preferring the early function obtained with knee disarticulation (132, 133, 142). Loder (145) examined 87 cases from the literature using the minimal requirements for a good result, as suggested by Jayakumar and Eilert (144), of acceptable gait, active knee motion of 10 to 80 degrees of flexion, varus/valgus instability <5 degrees, and no flexion contracture. He found that 53 of the 55 cases of Jones type Ia deficiency treated by Brown's procedure had a poor result because of flexion contracture. Many other authors have reported similar results and emphasize the need for strong active knee extension for a good result (132, 141–144). Simmons et al. were satisfied with the results from their evaluation of Brown's procedure (142). Their satisfaction was based more on the patients' feelings than objective assessment. Wada et al. (147) found that patients could weight bear in the home without assistive devices after the procedure but had poor function outside of the home and required a knee orthosis to ambulate. Like others, they

recommended against the Brown procedure in cases of complete absence of the tibia.

In patients with Jones type 1b tibial deficiency, it is important to monitor the patient over time, both for ossification of the proximal tibial remnant and to verify good active extension. It is possible that this remnant will be present, but good active extension will be absent or the remnant will not ossify. If there is active extension in a child, but the remnant is not sufficiently ossified by 1 year of age, the surgeon has several choices. The patient may be fitted with an extension-type prosthesis that accommodates the current foot position. Although this prosthesis (or prosthosis) is cosmetically unappealing, the child is usually quite functional. Alternatively, the surgeon may attempt to transfer the fibula to the unossified tibial segment with the goal of obtaining a successful tibial/fibular synostosis. Lastly, the physician can perform a Syme or modified Boyd amputation, fit with a prosthesis, and wait for ossification before performing the transfer.

In the rare patient with type Ia tibial deformity and proximal femoral deficiency with a very short limb, the best option may be to arthrodese the fibula to the distal end of the femur. The goal of this procedure is to increase the lever arm of the femoral segment, for the same reasons that a knee fusion is performed in children with PFFD.

In Jones type 2 deficiencies, the treatment of choice is to create a synostosis between the existing fibula and the tibial remnant to increase the length of the lever arm. A Syme or modified Boyd amputation, where the calcaneus is fused to the distal fibula instead of the tibia, is performed at the same time, and the patient is fit with a BK prosthesis (Fig. 30-24). Whether the synostosis is performed side-to-side or end-to-end, it is important to achieve good alignment of the fibula in relation to the knee joint. The residual proximal fibula should be removed to avoid problems later with prosthetic fit.

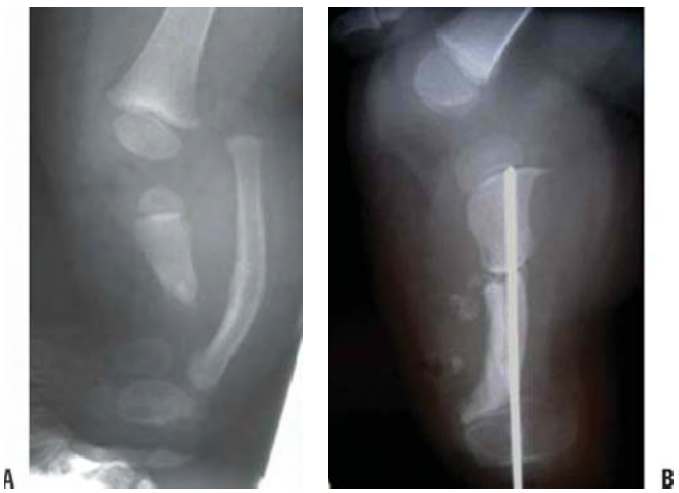


FIGURE 30-24. **A,B**: Pre- and postoperative lateral radiographs of a patient with type 2 tibial deficiency. The patient has undergone an end-to-end synostosis of the tibia and fibula with proximal fibular resection and a modified Boyd amputation with fusion of the calcaneus to the distal fibula.

Type 3 deficiencies of Jones are very unusual, and there is not much published experience. Jones et al. reported one case that was bilateral (148). They described a cartilaginous portion of the tibia, proximal to the ossified portion, which was "under voluntary muscle control." Their patient was treated with excision of the proximal fibula and Syme amputation. Fernandez-Palazzi et al. (127) had two cases in their report. Both were treated with Syme amputation, implying that there was an active quadriceps mechanism.

Jones type 4 deficiency presents a unique problem. At birth, the foot is deformed, often appearing like a clubfoot to the inexperienced. In addition, the amount of tibial shortening that will result is not apparent. This all makes it difficult for the parents to accept amputation. The difficulty for the surgeon is that this deformity is a spectrum of deformity. Garbarino et al. (149) have emphasized the distinction between a short tibia with a varus foot and a true congenital diastasis of the ankle joint. The former is usually amenable to reconstruction according to Schoenecker (131), whereas the true type 4 deficiency with diastasis of the ankle joint usually is treated with amputation (148, 150).

There are reports of reconstruction for the type 4 deficiencies, but in general the follow-up is short and the problems of a plantigrade foot and limb-length discrepancy are just beginning in these patients (149, 151–153). One patient followed up to the age of 15 years is described as having satisfactory ankle function and 6.5 cm of shortening (154), while another followed up to the age of 10 years (6 years after reconstructive surgery) is reported as having a stable ankle and plantigrade foot, but projected limb-length discrepancy is not mentioned (155). Choi et al. have recently reported on three patients treated with foot deformity correction with an Ilizarov device followed by distal tibial/fibular synostosis to stabilize the ankle mortise. This was followed with limb lengthening, including differential lengthening of the tibia and fibula. At skeletal maturity, all three had a plantigrade foot, less than a 3-cm leg-length discrepancy and were able to participate in group games or sports (156).

In their review of tibial deficiencies, Schoenecker et al. (130) reported on 10 patients with Jones type 4 deficiencies, of which nine had initial reconstruction of the foot. A Syme amputation was subsequently done in six of them, usually at the parents' request. Of the four patients who retained the foot, two had contralateral deficiencies in which the prosthesis accommodated the length discrepancy. One had a lengthening of 4.6 cm and one remained 4.8 cm short.

From the available information, it seems reasonable to attempt to retain the foot, if the deformity is at the less severe end of the spectrum, or if there is a significant contralateral deficiency. In most other cases, Syme amputation seems most reasonable.

Prosthetic Management. Depending on the severity of the anomaly and the surgery performed, there are several

prosthetic approaches to management. In children with type I tibial deficiency who have been treated with knee disarticulation and have a flare at the condyles, the prosthetic socket consists of a nonischial weight-bearing design with rotational control achieved through the intimate fit of the distal end of the socket over the femoral condyles and a well-formed gluteal impression. Suspension is usually achieved with the use of a segmented liner or bladder design that allows the wider condyles to pass through, while maintaining pressure over the femur just proximal to the condyles.

In cases in which the condyles are absent or there is the need to fit with a transfemoral socket, rotational control is achieved through proper contouring of the socket relative to the femur—the musculature surrounding the femur has a slight triangular shape in a cross-sectional view, with a flatter contour on the lateral surface, especially proximally. This allows a locking of the musculature which, with proper socket fit, decreases rotation. In addition, a silicone sleeve suspension may be used in conjunction with a pull-through strap to secure the liner. If all other procedures fail, a standard Silesian belt (around the pelvis) may be utilized. The total elastic suspension (TES) belt offers excellent suspension and flexibility of form, and it aids in control of the prosthesis. However, the Silesian belt and TES will interfere with grooming and toilet training.

In the knee disarticulation (or transfemoral) prosthesis for children, there are differences of opinion as to when young children are able to handle an articulated knee. Traditional established practice is to first fit the child with a locked knee and allow an articulating knee at approximately 3 to 5 years of age. In contrast, Wilk et al. (157) advocate the use of articulating knees in children as young as 17 months. Children as young as 11 months can be appropriate candidates for articulated knees (155). The children learn how to handle the knee very quickly, and there is very little need for any type of device to temporarily stabilize the knee. The use of a knee joint at this stage permits more normal development, allowing bent-knee sitting, side sitting, crawling and kneeling on hands and knees, and easier pull to a stand. With a pediatric knee, children can reduce or eliminate a circumducted gait pattern.

In type II cases, in which a tibial segment has been preserved or the fibula has been joined to the tibial remnant, a modified transtibial prosthesis or a Syme prosthesis is utilized. Unlike the standard transtibial design, the socket will incorporate SC and suprapatellar proximal brim lines that will aid in the control and stability of the knee and prevention of a hyperextension moment, respectively. In some instances in which knee stability is less than optimal, outside joints and a thigh cuff or lacer may be required. These are used as a last resort and often contribute to increased weakening of the musculature as a trade-off for increased control and alignment.

Authors Preferred Recommendations. For patients with no active knee extension and Jones type Ia tibial deficiency (complete absence), the authors recommend knee disarticulation and prosthetic fitting. The authors have not seen any patients as described in the literature with complete tibial absence yet the presence of adequate active knee extension, but would recommend knee disarticulation and prosthetic fitting for these patients as well because of its high functional result compared to the poor functional results reported for the Brown procedure. For patients with some active knee extension and Jones type 1b or type 2 tibial deficiency, the authors recommend waiting for the tibial remnant to ossify, then performing a tibial–fibular synostosis in an end-to-end fashion. At the same time as the synostosis, a modified Boyd amputation is performed, with fusion of the distal fibula to the calcaneus. If the proximal fibula is proximally displaced, prominent, and if the knee has varus deformity or instability, resection of the proximal fibula is recommended as well.

Timing of the tibial–fibular synostosis, modified Boyd amputation, and possible proximal fibular resection is undertaken at approximately 1 year of age unless the proximal tibia is unossified. The authors recommend fitting the child with an unossified proximal tibia with an extension prosthosis that accommodates the foot deformity and waiting until the proximal tibia ossifies. This has the benefit of one definitive surgical episode while allowing the child to walk at a normal developmental age and has the added benefit of saving the toes for possible transfer to the hand if hand anomalies coexist.

The authors have no experience with Jones type 3 tibial deficiency but agree with Schoenecker that ankle disarticulation and prosthetic fitting are appropriate. For Jones type 4 cases and a projected limb-length discrepancy of 5 cm or less, the authors recommend early soft-tissue correction of the foot deformity with later contralateral epiphysiodesis to achieve limb-length equality. For those cases with a projected discrepancy above 5 cm, Syme amputation and prosthetic fitting is preferred.

Pearls/Pitfalls. As mentioned previously, outcomes of fibular centralization are poor. The literature suggests that this almost uniformly results in a poor functional result and subsequent knee disarticulation. Initial knee disarticulation in patients without active knee extension results in less surgery and a more functional result.

During tibiofibular synostosis surgery, a few points are worth mentioning. The proximal fibula in these patients often is proximally displaced and prominent laterally. The surgeon should consider resection of this at the time of tibiofibular synostosis surgery so it does not cause difficulty with prosthetic wear in the future. With regard to the technique of synostosis, the authors have found that end-to-end apposition of the tibia and fibula results in superior lower limb alignment for prosthetic fitting. The fibula usually needs to be slightly shortened to take tension off of the soft-tissue structures to achieve this alignment, which is of no consequence.

Complications. The complications of Syme versus Boyd amputation were previously discussed in the section on fibular deficiency.

In patients planned to have a tibiofibular synostosis, nonunion can occur, particularly if the tibial segment is unossified. The authors suggest waiting for tibial ossification before attempting synostosis, even if it delays the achievement of normal motor milestones for the child.

Just as progressive foot deformity can occur with tibial lengthening in fibular deficiency, worsening of foot deformity can occur after previous correction of the foot in type 4 tibial deficiency. Stabilizing the ankle mortise with a distal tibial/fibular synostosis and including the foot in the Ilizarov frame during lengthening conceptually should reduce this complication. Another solution is to fuse the distal fibula to the posterior facet of the calcaneus, thereby stabilizing the foot under the fibula (147).

Femoral Deficiency

Definition and Classification. The term "femoral deficiency" encompasses a wide-spectrum pathology, varying from a short but relatively normal femur to almost total absence of the femur with only the distal femoral condyles present. Aitken popularized the term "proximal femoral focal deficiency" to refer to the more severe forms of this disease, although the deficiency is not isolated to the proximal femur and is often part of a global limb deficiency.

There have been numerous classifications of PFFD (32, 34, 73, 158–162). Some systems are radiographic in nature, while others are treatment based. For those that are treatment based, classification groups are slightly different, which reflect the differing opinions on optimal treatment. No classification system adequately describes the whole spectrum of the limb deficiency, which includes both bone and soft-tissue anomalies.

The Aitken classification is the most widely used system (158). It is a radiographic classification system based on the severity of the radiographic findings of the hip and femur. As is true with all radiographic classification systems where there is temporal evolution of the radiographic findings, the classification of a particular patient may change over time as the radiographs evolve. Goddard et al. suggested that the Aitken classification was easier to apply to a radiograph taken at 12 to 15 months of age, when the child is starting to stand and walk, rather than one taken immediately after birth (163).

In class A, the femur is short, with the most proximal ossified portion at or slightly above the acetabulum [Fig. 30-25: (Right)]. There is incomplete ossification in the subtrochanteric region of the femur which will ossify over time. There is often a subtrochanteric varus deformity with ossification. The femoral head may not be visible on radiographs, but there is a well-formed acetabulum that implies its presence.

In class B, there is a more extensive defect or absence of the proximal femur (Fig. 30-26). The acetabulum shows more signs of dysplasia than in type A. In addition, the most proximal part of the femur is part of the femoral shaft, which is located above the level of the acetabulum. The radiographic defect in the subtrochanteric region, in contrast to class A, does not ossify with time and instead forms a pseudarthrosis.

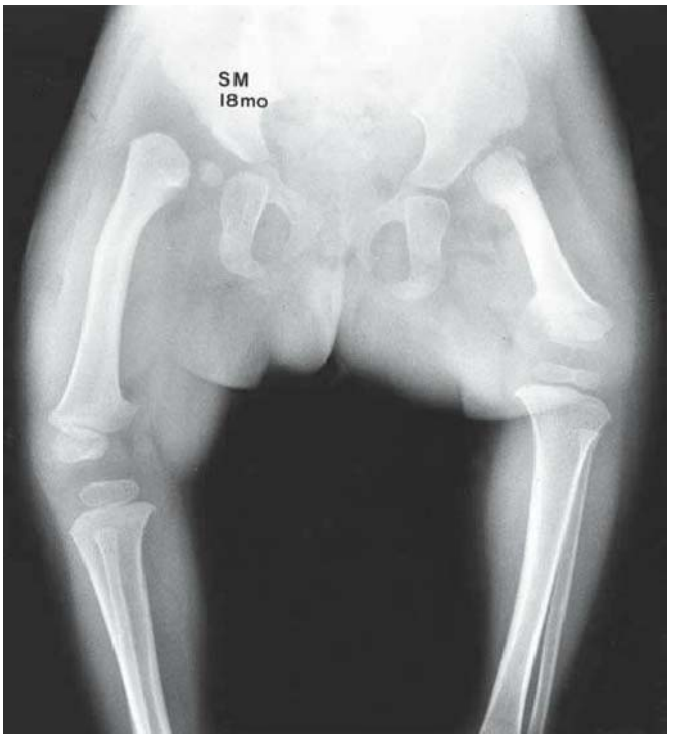


FIGURE 30-25. AP pelvis of an 18-month-old child with bilateral PFFD. The right hip is an Aitken class A and demonstrates the presence of the ossific nucleus and a good acetabulum. The femoral metaphysis lies above the level of the ossific nucleus. There is a cartilaginous connection between the metaphysis and the femoral head, which will usually ossify by skeletal maturity, but often with a significant varus deformity. The opposite hip is an Aitken class C PFFD. This patient demonstrates the difficulty with limb-length difference in some patients with bilateral PFFD.

In Aitken class C, the acetabulum is severely dysplastic [Fig. 30-25 (left)] and (Fig. 30-27), signifying that the femoral head is absent and will not ossify with time. The femoral shaft is shorter than in class B, and the entire proximal femur does not ossify with time.

In Aitken class D, the femoral shaft is essentially absent (Fig. 30-28). The distal femoral condyles are seen at the level of where the acetabulum should be. The lateral pelvic wall is flat, without any evidence of acetabular development.

Gillespie (134) proposed a three-tiered classification system based on treatment recommendations rather than radiographs. Group A are those with congenital short femur indicated by clinical hip stability, lack of significant knee-flexion contracture, and the foot of the affected extremity lying at or below the midpoint of the opposite tibia (Fig. 30-29). These patients have a hip that is stable to weight bearing and a femoral segment that is >60% of the contralateral side. These patients are likely candidates for limb lengthening. His group B patients include patients with a hip that is unstable with weightbearing and have a femoral

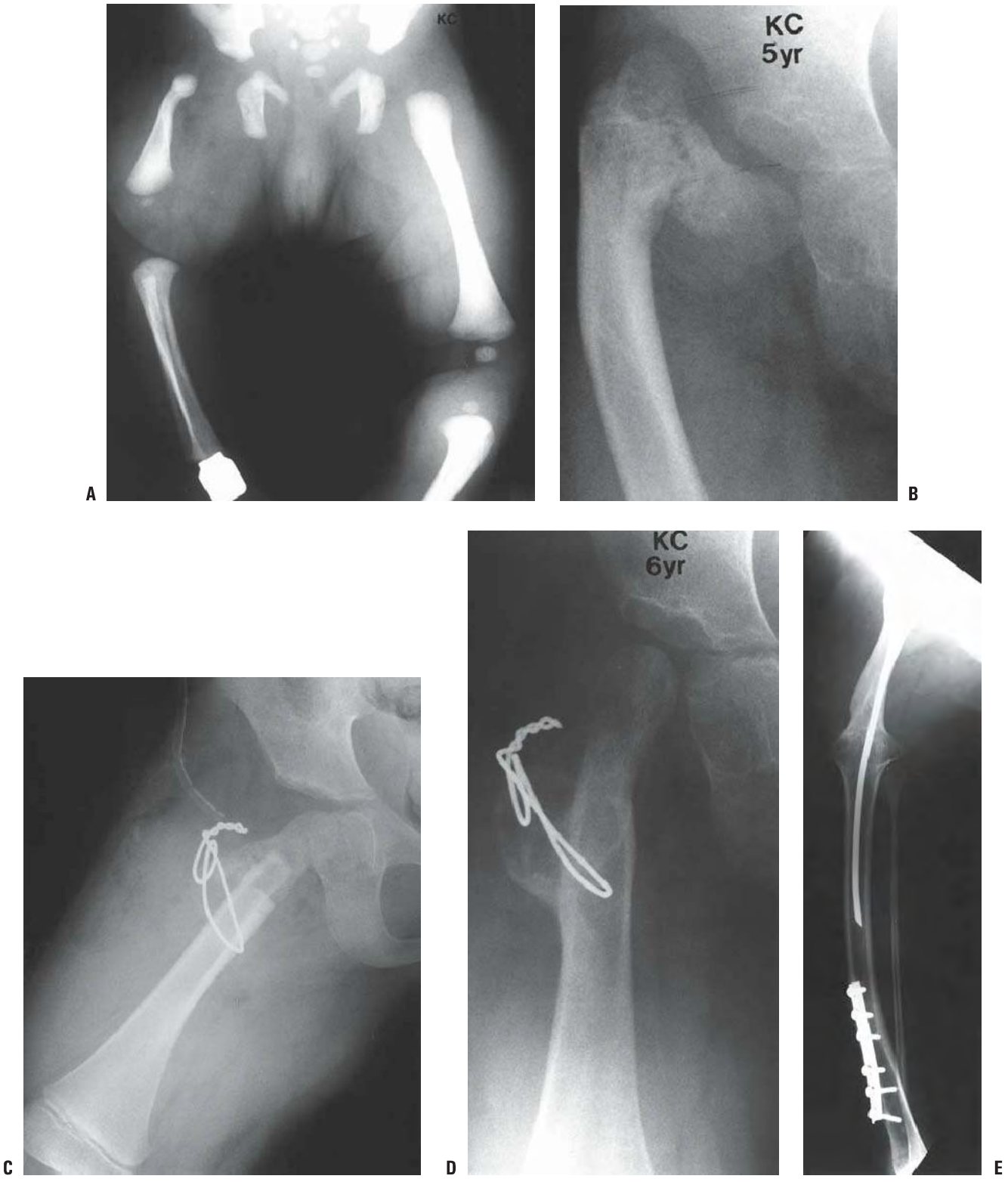


FIGURE 30-26. **A**: AP radiograph of the pelvis and limbs of a newborn girl with Aitken class B PFFD. Note the short femoral segment and the well-developed acetabulum, although the femoral head is not visible. **B**: By 5 years of age, the femoral head is ossified and the cartilaginous connection between the femoral head and the subtrochanteric region of the femur has undergone considerable ossification. However, a pseudarthrosis persists and a significant varus deformity has developed. **C**: The femur after correction of the varus with a spike type of osteotomy. **D**: The result 1 year later. Now faced with a projected discrepancy of 20 cm, the parents elect a Van Nes rotationplasty. This was done with part of the rotation through the knee arthrodesis, and the remainder through the tibia. **E**: The radiographic result. The patient had one additional derotation performed through the midtibia at the age of 10 years.

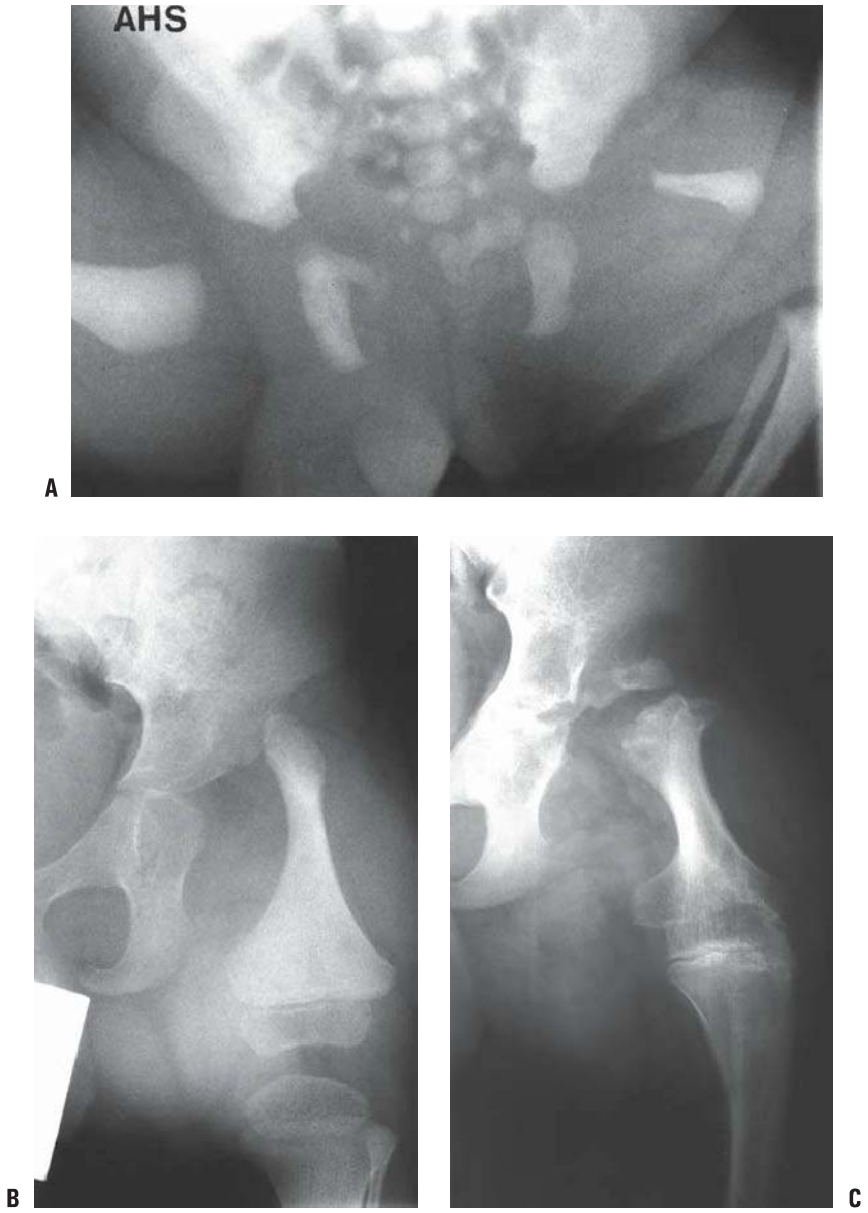


FIGURE 30-27. The AP pelvis and limbs of a newborn boy **(A)** and at 3 years of age, just before surgery **(B)**, with Aitken class C PFFD. Note the very short femoral segment and the lack of acetabular development. The same patient is seen in **(C)** at the age of 12, following a Syme amputation and knee arthrodesis with preservation of the proximal tibial physis. There is still no appearance of a proximal femoral ossific nucleus.

segment that is <50% of the contralateral side (Fig. 30-30). Group C patients have only a small tuft of distal femur present and no acetabular development (Fig. 30-31). He recommends prosthetic treatment for his group B and C patients.

Paley based his classification on treatment recommendations as well, with a special emphasis on what is necessary for limb lengthening and reconstruction (164). He emphasized the importance of the degree of dysplasia and function of the knee for a good outcome with lengthening. His type 1 is similar to Gillespie's group A but is divided into three subgroups depending on problems at the hip and knee which will have to be addressed either before or at the same time as lengthening. Type 2 has a mobile pseudarthrosis with or without a mobile femoral head. Stabilization of the pseudarthrosis or of the proximal femur in relation to the pelvis is an essential prerequisite of lengthening. When the femoral head is immobile or absent, stabilization of the external fixator to the pelvis is necessary, frequently combined with a valgus extension proximal femoral osteotomy. Type 3 is similar to Gillespie's group C. If knee motion is <45 degrees, functional gains with lengthening are doubtful.

An unusual variant of PFFD is that seen with a bifurcated distal femur. On radiographs, the femur has the shape of an inverted "Y" (165). In addition, these patients always have complete absence of the tibia and often exhibit hand

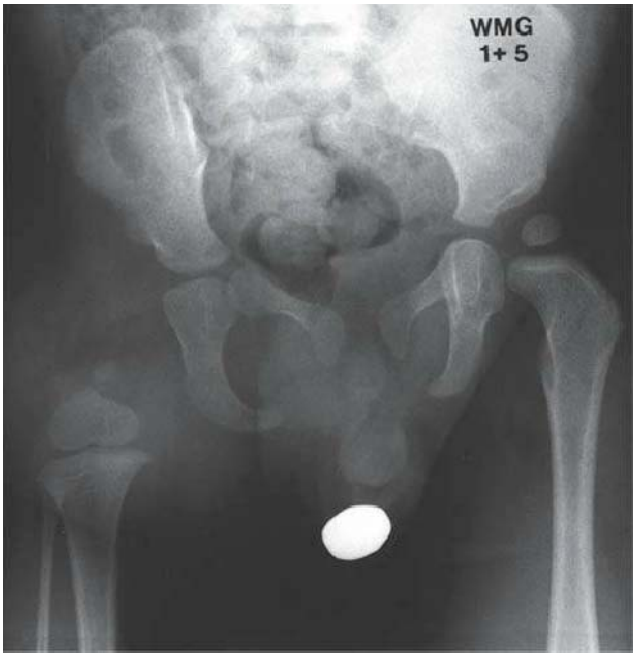


FIGURE 30-28. AP radiograph of the pelvis and femur of a boy aged 1 year and 5 months with Aitken class D proximal focal deficiency. There is little femur present, and no sign of acetabular development. He underwent knee arthrodesis and Syme amputation at 2 1/2 years of age.

FIGURE 30-30. Clinical photograph of a patient with Gillespie type B PFFD. The femur is <50% of the length of the contralateral side. In addition, the proximal femur is unstable with weight bearing.

FIGURE 30-29. Clinical photograph of a patient with Gillespie type A PFFD. Note the lack of significant knee-flexion contracture on the affected side, with the foot falling below the midpoint of the contralateral tibia.

FIGURE 30-31. Clinical photograph of a patient with Gillespie type C PFFD. There is severe shortening of the femur, and there is complete absence of a femoral–acetabular articulation.

and foot anomalies as part of the tibial deficiency ectrodactyly syndrome, which is inherited as an autosomal dominant trait (166). Treatment in these cases is knee disarticulation and removal of the segment of distal femur in poorest alignment.

Etiology and Epidemiology. Femoral deficiencies are one of the teratogenic effects of thalidomide. In the modern age, femoral deficiencies are sporadic, unless they are part of a larger syndrome such as femoral hypoplasia–unusual facies syndrome, which exhibits an autosomal dominant inheritance pattern.

Clinical Features. The appearance of patients with femoral deficiency is classic and should be easily recognized. It will be bilateral in 15% of the cases. The femoral segment is short, flexed, abducted, and externally rotated. The hip and knee joints exhibit flexion contractures. The proximal thigh is bulbous and rapidly tapers to the knee joint (Fig. 30-32). Fibular deficiencies are so common in association with PFFD that the valgus foot and other characteristics of fibular deficiency are almost a part of PFFD. PFFD is associated with fibular deficiency in 70% to 80% of cases (167). In addition, approximately 50% of the patients will have anomalies involving other limbs (158, 167).

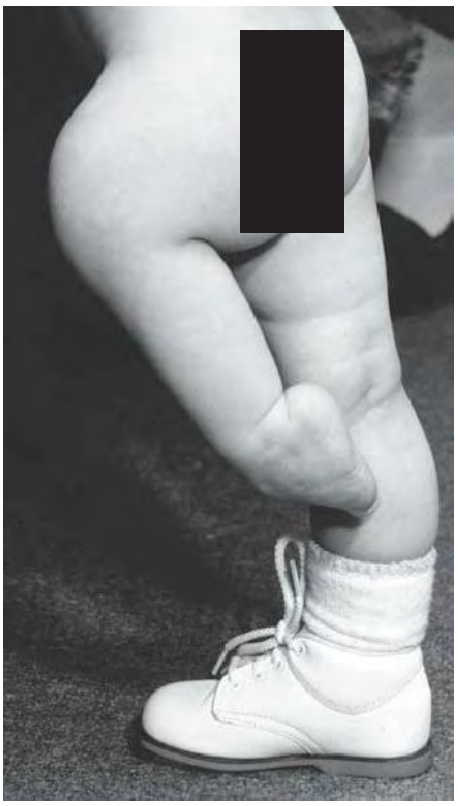


FIGURE 30-32. This photo of a 12-month-old girl who is pulling to the standing position demonstrates the clinical features of PFFD: a very short and bulbous femoral segment, which is flexed, abducted, and externally rotated.

Examination of the hip joint is difficult because of the bulbous thigh and short femoral segment. Pistoning may be apparent because of associated hip instability. The knee is always unstable in the AP direction.

Radiographic Features. Most of the radiographic features were covered in the description of the Aitken classification. In patients with a congenital short femur, the only finding may be slight coxa vara and an anterolateral bow in the femoral shaft. In addition, the findings of fibular deficiency are often evident, as up to 50% of these patients have concurrent fibular deficiency.

Other Imaging Studies. Recently, an MRI classification of femoral deficiency has emerged, which partially addresses the difficulty of the unossified proximal femur on plain radiographs (168). The authors demonstrated that the cartilage anlage in the proximal femur region was well-visualized with MRI and correlated to the plain radiographic appearance over time to the initial MRI. They suggest that an MRI classification should be used in lieu of a plain radiographic classification.

Pathoanatomy. There are few anatomical studies of patients with femoral deficiency. Most of what we know about the pathoanatomy are based on imaging studies. There are pathologic changes throughout the entire limb of varying severity. The acetabulum can exhibit mild dysplasia and retroversion in mild cases, and it can essentially be absent in severe cases. The proximal femur can have delayed ossification and a varus deformity in the intertrochanteric region or there can be a pseudarthrosis. In severe forms, it is completely absent. With regard to the knee, findings can range from mild anterior/posterior laxity to complete absence of the cruciate ligaments to severe flexion contracture. As mentioned previously, the lower leg can be normal, but often exhibits fibular deficiency, possibly with severe foot deficiencies that occasionally go along with that disease process.

Pirani et al. (169) recently described the MRI appearance of the musculature around the hip and proximal femur. Most muscles were hypoplastic, except for the obturator externus and the sartorius, which were hypertrophied. In addition, the obturator externus coursed in an abnormal direction in more severe cases. In describing their MRI classification for PFFD, Maldjian et al. (168) showed, in patients who do not develop pseudarthrosis of the proximal femur, that there is a cartilaginous anlage that attaches the proximal femur to the femoral head.

Natural History. Regardless of the severity of the femoral deficiency, children will usually walk at the normal developmental age. Children with a mild congenital short femur walk with a slight Trendelenburg gait and have a mild limb-length discrepancy. They often compensate with varying degrees of hip, knee, and ankle flexion on the contralateral side if

untreated. Children with more severe forms often choose to walk on the knee of the unaffected side to make up for the severe limb-length discrepancy during the first few years of life until the discrepancy is severe and treatment is sought.

Treatment Recommendations

Nonsurgical Treatment. Most patients will benefit from a treatment plan that includes surgical intervention, be it lengthening, amputation, or rotationplasty. However, there are few reports that demonstrate patient-perceived outcomes of amputation and prosthetic fitting versus accommodative prosthetic fitting without amputation have similar function results (170). Thus, accommodative prosthetic fitting might be an option for the rare patient who does not want to undergo any surgical intervention. More often, however, prosthetic fitting is a bridge treatment instituted when the child is ready to walk until age 2 1/2 to 3 years of age, when definitive treatment is planned. This prosthesis is nonconventional in design and is sometimes referred to as an extension prosthesis or a "prosthosis." It is designed to fit the extremity without any surgical modification to it (Fig, 30-33). The flexion, abduction, and external rotation of the proximal segment (the femur) are accommodated in the alignment. At this young age, the knee joint of the prosthesis can be omitted.

The treatment of children with bilateral PFFD is predominantly nonoperative. These children do not use prostheses. Occasionally, if there is asymmetric involvement of the limbs, a limb-length discrepancy can exist. Treatment is individualized to each patient.

Surgical Treatment. Surgical treatment aims to compensate for the functional problems the patient experiences. The most obvious of these is the shortening of the limb. Less obvious is the problem with hip function and its relation to the alignment of the limb. Because of the flexed and externally rotated femoral segment, the knee remains flexed, and the leg and foot are anterior and lateral to the axis of the body (Figs. 30-32 and 30-33). Without surgical treatment, the patient must lean laterally and posteriorly during stance phase on the affected limb to move the weight-bearing line so that the proximal femoral segment will be more stable. This gait pattern is accentuated because of the additional muscle deficiency around the hip. The knee will have varying degrees of instability. The function of the foot will depend on the severity of any associated deficiencies of the leg, for example, fibular deficiency.

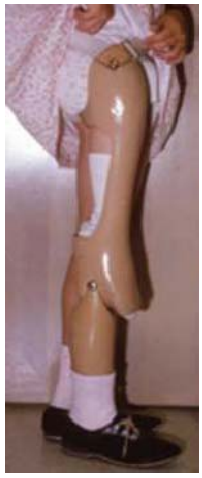


FIGURE 30-33. Lateral photograph of a patient with an extension prosthesis that accommodates the retained foot proximal to the terminal end of the prosthesis. This type of prosthesis can be useful if the parents refuse foot removal or in the young toddler before definitive treatment.

There are three main treatment strategies for PFFD patients; knee fusion with foot ablation, Van Nes rotationplasty with knee fusion, and limb lengthening. Each strategy is vastly different from the other, and early decision making is necessary to put the child on the proper path. Fortunately, most of these decisions can be postponed until 2 1/2 to 3 years of age, because this is the best age to perform these surgical options.

Most authors suggest limb lengthening if the predicted discrepancy at maturity is <20 cm; the hip is, or can be, made stable; and there is good knee, ankle, and foot stability and motion. Such cases require multiple-staged lengthenings in addition to a contralateral epiphysiodesis and sometimes a shoe lift. The timing and staging of these procedures depends on the choice of the physician, but will usually not start before the age of 3 years. Reports of the functional outcome in patients followed up to maturity and into adulthood are lacking.

If the discrepancy is predicted to be >20 cm at maturity, or for any other reason lengthening is not chosen as a treatment, a decision should be reached about the best approach to prosthetic fitting. Surgery can make the residual limb a more efficient lever arm to power the prosthesis. In addition, foot ablation can lead to a more cosmetic appearance of the prosthesis.

Knee Arthrodesis. Arthrodesis of the knee joint is a standard procedure in children with PFFD undergoing prosthetic fitting. It creates a single, longer, and more efficient lever arm, which is easier to contain within the prosthesis. This will greatly enhance prosthetic function and reduce energy consumption. The proximal femoral segment deformity (flexion, abduction, and external rotation) does not need to be compensated for at the time of knee fusion. If the tibia is fused in line with the femur, subsequent ambulation with a prosthesis will gradually correct the soft-tissue balance around the hip and realign the limb with the contralateral side.

Depending on the length of the femoral segment and the limb as a whole, it is usually desirable to remove at least one of the growth plates at the knee at the time of fusion. This is usually the case in Aitken class A, B, and C deformities. Without removing at least one of the epiphyses and physes at the knee, the limb will be too long (Fig. 30-34). The reason for this is that most above-knee prosthesis designs need approximately 7 cm to accommodate a prosthetic knee joint. If the ipsilateral lower leg segment is normal in length or mildly shortened, as it often is, then the growth of the lower leg segment alone will

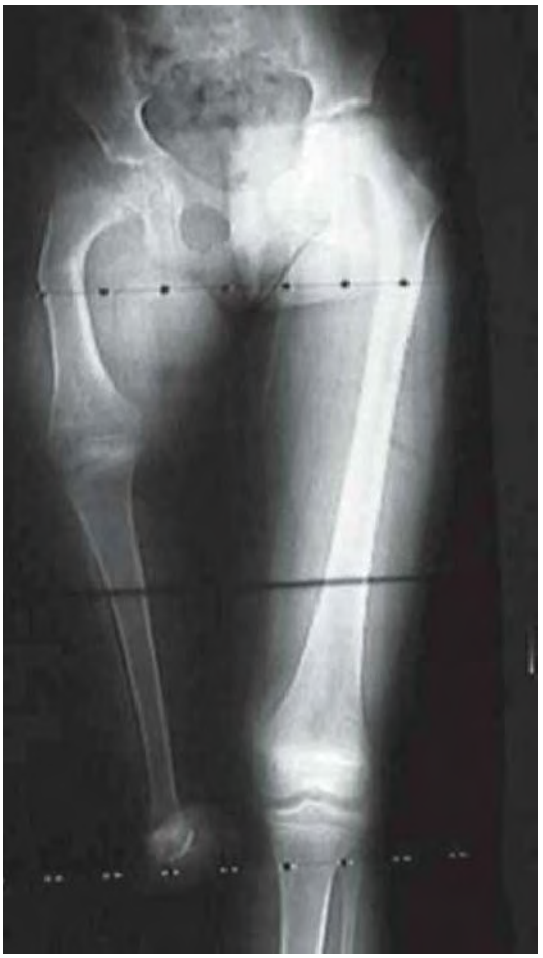


FIGURE 30-34. An AP standing lower extremity radiograph of a patient with PFFD after knee fusion. Neither the distal femoral or proximal tibial epiphyses were removed at the time of fusion, resulting in a residual limb that falls below the contralateral knee. Coupled with the length required to fit a prosthetic knee joint (typically >7.5 cm), this will result in a residual limb that is significantly too long for the patient. Removal of the distal femoral epiphysis and physis at the time of knee fusion is indicated except when the residual femur is extremely short and the tibia is significantly short as well.

usually give enough limb length for successful prosthetic fitting. Any additional length contributed by the femur is unnecessary and interferes with accommodating the knee joint in the prosthesis. The author's experience is that, in considering patients with PFFD with or without fibular hemimelia, 90% of children had at least 90% of the normal length of the tibial segment. In the children where this was true, patients at the time of knee fusion underwent excision of both distal femoral and proximal tibial epiphyses and physes, and no residual limb was too short to successfully fit with an above-knee prosthesis. In some cases, with more pronounced tibial and femoral shortening, it may be advisable to remove neither or one epiphysis and physis (171). Calculation of the anticipated length of both limbs at maturity by means of the Green-Anderson growth charts (Tables 30.2 and 30.3), as described earlier, will help with the answer.

Fixation at the fusion site is often a rigid intramedullary rod inserted from the proximal femur across the fusion site into the distal segment, with an additional K-wire across the fusion site to give the construct rotational stability. The patella may or may not be excised during fusion. If it is excised, the bone can be used as graft at the fusion site. There have been a few cases reported of late-onset patellofemoral pain and arthritis on radiographs in PFFD patients with knee fusions and retained patellae (172). After surgery, limb stability is enhanced with a spica cast. The patient is usually ready for prosthetic fitting in 6 weeks and for ambulation as soon as the prosthesis is ready.

Amputation of the Foot. With the knee fused, ablation of the foot is desirable in most situations. One reason is to ensure the residual limb will be short enough to accommodate an internal knee joint when the child is older. The other reason is that it becomes increasingly difficult to fit the growing foot in a cosmetically acceptable socket. Whether or not a Boyd or Syme amputation is performed is largely surgeon dependent, and the relative merits of each procedure have been previously discussed.

Van Nes Rotationplasty. In the Van Nes rotationplasty, the limb is rotated 180 degrees, predominantly through the knee arthrodesis, with some additional rotation through the tibia if necessary. The goal is to have the ankle/hindfoot complex of the short limb at the level of the knee on the long limb at maturity. The foot now functions like the residual tibia in a below-knee amputation, thereby allowing the patient to function more like a BK amputee than one with a knee disarticulation (Figs. 30-35A–D). Sufficient ankle and hindfoot flexion and extension, as well as ankle stability and alignment, are necessary for this type of treatment.

The rotationplasty was first described in 1930 by Borggreve (173), for acquired traumatic limb-length discrepancy. Van Nes (174) later used the procedure for three cases of congenital deficiency of the femur. Initial reports of rotationplasty for treatment of PFFD by Kostuik et al. (175) and Torode and Gillespie (176) have been followed by more recent reports by Friscia et al. (177) and Alman et al. (178).

The main complication of the procedure is either failure to achieve sufficient rotation at surgery or subsequent derotation with growth. Kostuik et al. (175) recommended waiting to perform the surgery until the child was older. However, this prevents the child from deriving the gait benefits for several years. Subsequent reports have not found this to be so great a problem. Also, derotation can be treated with revision surgery.

Even though the functional results of the surgery are superior to that of an above-knee prosthesis wearer, parents and physicians are sometimes reluctant to perform it because of the cosmetic appearance of the foot pointing backward. It appears, however, that this problem is overrated by medical staff, compared to the patients themselves. Alman et al. (178) found no difference in the perceived physical appearance of children treated with rotationplasty, compared to knee

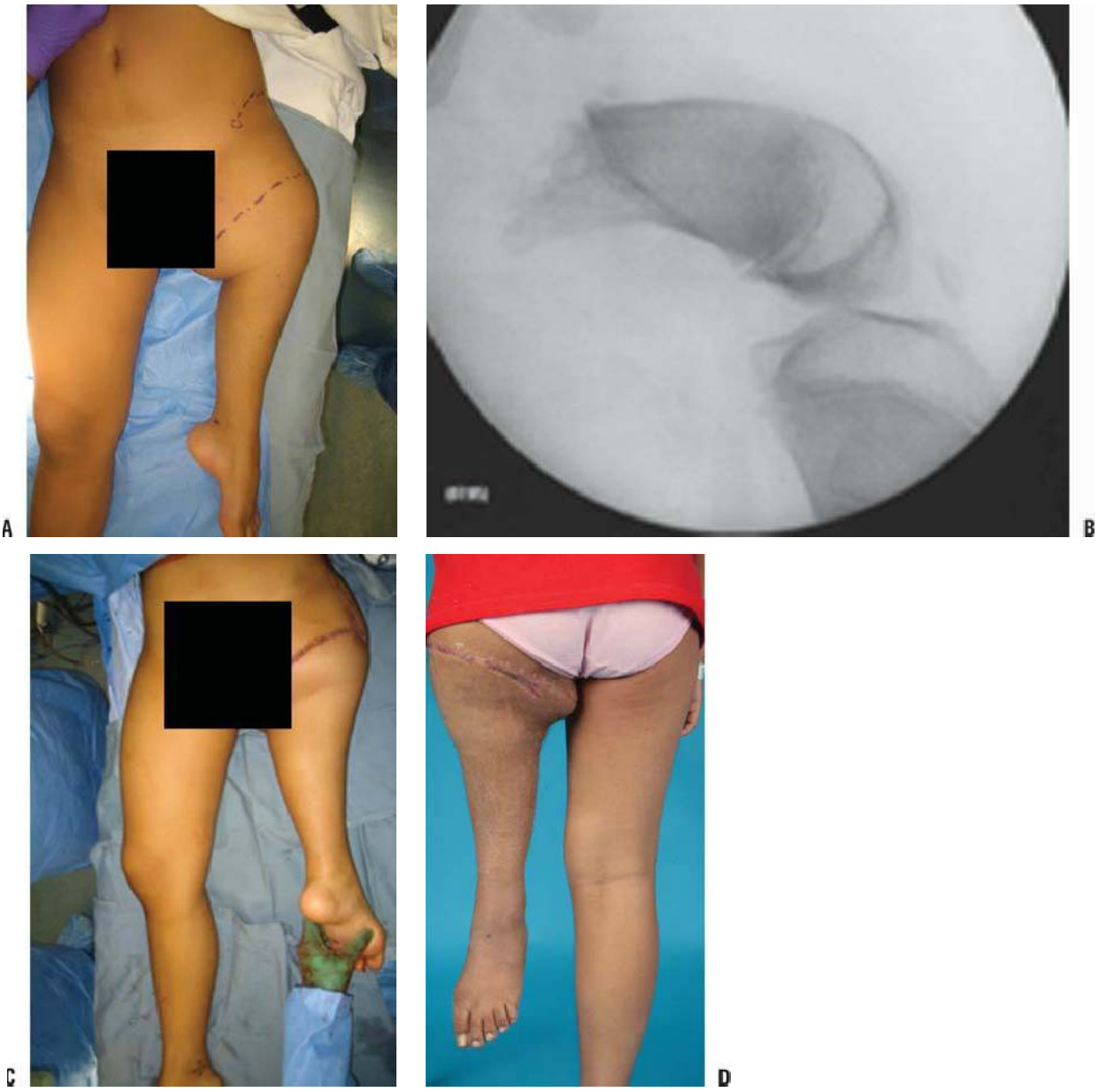


FIGURE 30-35. **A**: Preoperative clinical photograph of a patient with Gillespie type 2 PFFD. **B**: An intraoperative fluoroscopic radiograph demonstrating the extreme shortening of the femoral segment and lack of any acetabular development. **C**: Postoperative clinical photograph after Van Nes rotationplasty. **D**: Postoperatie standing clinical photograph at 1 year of follow-up.

arthrodesis and Syme amputation. In the report of Friscia et al. (177), one patient subsequently had a Syme amputation at the parents' request. Two recent studies evaluating the quality of life in patients who had rotationplasty for sarcoma treatment demonstrated that although physical function was less than that in healthy peers, psychosocial adaptation and life contentment were about the same (179, 180). This emphasizes the importance of proper presurgical preparation of the parents and of the patient, if she or he is old enough. This is best accomplished by seeing other patients with a rotationplasty, along with the use of videos of patients, teaching dolls, and so on.

It is imperative that the ankle must be sufficiently normal to serve as a knee. This is particularly important to determine, because up to 70% of children with PFFD will also have a fibular deficiency on the same side. Although some valgus alignment of the foot and ankle can be compensated for in the prosthetic alignment, the deformity may progress with age. Severe valgus and equinus deformities, with a deficient foot, are contraindications to the procedure.

Additional preoperative preparation includes toe, ankle, and hindfoot strengthening, in particular, because these are the structures that will power the new knee joint. Equinus position should be emphasized, because this will place the

foot in the best mechanical position. Children who have mild equinus contractures of 30 degrees or less will usually stretch these out with prosthetic use and do not need special attention preoperatively. Crutch training should be done preoperatively, as in all elective surgery that will require crutch use postoperatively.

The improvement in function with the rotationplasty, compared to other procedures, has been documented both for patients with tumor (181–183) and for those with PFFD (178, 184). These studies demonstrate patients with rotationplasty function better than those with knee arthrodesis and foot ablation, not quite so well as those with a below-knee amputation, and not as well as those who have rotationplasty for noncongenital conditions, for example, tumor. Those with rotationplasty for noncongenital conditions probably do better because of the normal hip function that remains one of the major problems in those with PFFD.

Limb Lengthening. The general concepts surrounding limb lengthening are covered in other chapters. Several issues specific to limb lengthening in patients with congenital short femur are worthy of discussion. Issues concerning knee instability during lengthening were previously discussed in the section on fibular longitudinal deficiency.

Stabilization of the Hip. Most patients with PFFD, whether undergoing lengthening or prosthetic fitting, will have hip instability. This is not only because of the deficient bony anatomy, but also because of the deficient musculature. This has resulted in some controversy about the value of surgical procedures to stabilize the hip. Some feel that nothing of functional value is gained and surgical intervention is not warranted (165, 171, 185), whereas others feel that surgical correction can be of value (32, 163, 186, 187). It is the authors' opinion that in Aitken class A and B patients who have a mobile femoral head within the acetabulum, surgical correction of an existing pseudarthrosis with correction of the varus and retroversion deformity if there is less than a 110 degree neck-shaft angle is beneficial (Fig. 30-26B–D).

There are multiple anatomic problems to consider: the pseudarthrosis and consequent malalignment, the flexion/abduction/external rotation soft-tissue contracture, and the bony stability of the femoral–pelvic articulation. In those patients for whom lengthening is planned, it is necessary to obtain good containment of the femoral head, which may require an acetabular procedure. In contrast to the typical anterolateral acetabular deficiency as seen in DDH, the acetabulum is often retroverted in PFFD, resulting in a lack of posterior and lateral coverage (188). Therefore, reshaping acetabular procedures must address this posterior deficiency. In addition, femoral retroversion and varus are also usually present and should be corrected prior to lengthening. The soft-tissue contractures include the hip flexors (predominantly the rectus femoris and iliopsoas) and hip abductors (primarily the gluteus medius and minimus). These may be addressed as part of the proximal femoral reconstruction as described by Paley et al. Alternatively, the proximal femoral reconstruction may be performed without addressing the soft-tissue contractures. Anecdotally, the soft-tissue contractures may stretch out over time with prosthetic use, although there is insufficient literature to advocate one approach over another.

In patients with Aitken class B PFFD, there will be a pseudarthrosis of the femoral neck. This can be repaired while, at the same time, restoring more normal alignment. It may not be necessary to wait until complete ossification of the femoral neck to perform this procedure (189). Ossification may accelerate after realignment.

Iliofemoral Arthrodesis. There are two types of iliofemoral arthrodeses described. These procedures are an attempt to address the problem of hip instability predominantly in patients with Aitken class D femoral deficiency.

In 1987, Steel (190) described arthrodesis of the distal femoral segment to the pelvis in the region of the acetabulum in four patients. The femur was fused in 90 degrees of flexion so that it was perpendicular to the axis of the body. This results in knee extension being equivalent to hip flexion, and knee flexion being equivalent to hip extension.

More recently, Brown (191) has described a rotationplasty in conjunction with iliofemoral arthrodesis. In this procedure, the distal end of the femur is rotated 180 degrees before it is joined to the ilium with its axis in line with that of the body. The knee now functions as the hip joint, and the ankle now functions as the knee joint, as in a Van Nes rotationplasty (Fig. 30-36). In his opinion, the complication of derotation was less likely in these patients than with the Van Nes procedure.

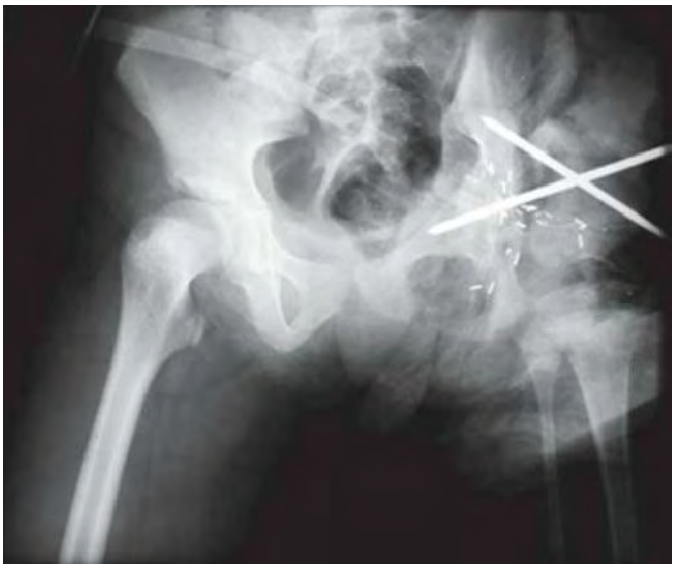


FIGURE 30-36. AP pelvis radiograph of a patient with PFFD who underwent 180 degree rotation and fusion of the proximal femoral segment to the ilium (as described by Brown KL. Resection, rotationplasty, and femoropelvic arthrodesis in severe congenital femoral deficiency. A report of the surgical technique and three cases).

Both of these procedures have had limited use. There are significant problems in achieving an arthrodesis, and the distal femoral segment cannot be allowed to grow too long. Additional surgical procedures are to be expected. As yet, there are only very limited reports on the functional advantages (190, 191).

Prosthetic Management. Initial prosthetic management of the child with PFFD begins with fabrication of an extension or nonstandard prosthesis, with or without an activated knee joint (Fig. 30-33). With the foot positioned in plantar flexion, the limb is cast proximal to the hip joint, and the prosthesis fabricated with a prosthetic foot positioned under the shortened limb. The ischial containment socket has been called a "ship's funnel" because of the resemblance to the engine air intake funnels of ocean vessels. This drastic socket design is necessary because of the flexed hip and knee that must be contained within the socket while attempting to gain ischial support.

The purpose of the extension-type prosthesis is to equalize limb length, in preparation for early ambulation, while affording time for surgical decisions. There are four indications that have been identified relating to the fitting of nonstandard prostheses (192):

1. When the patient is still too young for surgical conversion.
2. When the patient or parent refuses surgical intervention, and a prosthesis is necessary for ambulation.
3. In bilateral cases, when extra height or better balance is the goal.
4. When there is lower extremity involvement, combined with bilateral upper extremity absence, requiring the feet for ADL.

When foot ablation with knee fusion option is chosen, the prosthesis resembles a knee disarticulation prosthesis, except for the need for ischial weight bearing and high lateral brim containment to aid in hip stability. Weight bearing is divided between the ischium and the distal heel pad. Full distal weight bearing could severely compromise hip function over a period of time, because of the inherent instability of the hip with possible proximal migration of the femur. Prosthetically, fusion of the knee with correction of the angular deformities results in improved gait and ease of fitting because of the single skeletal lever arm (193). During growth, the child should be evaluated periodically for the relative length of the two limbs so that, if needed, distal femoral epiphysiodesis can be performed. This will allow fitting of an optimal knee joint when the patient is fully grown while maintaining the knees at the same level.

In the small child, and when the residual limb is longer than the opposite femoral segment, external knee joints may be used. As the child grows, an internal four-bar knee can be used. More about the indications and selection of knee joints is discussed later in this chapter.

A foot amputation without knee fusion results in difficulty with prosthetic management. Movement within the prosthesis, at the level of the anatomic knee, and the increased need for an intimately fitted socket, foster a decreased stride length and increased pelvic movement. However, in the child with an Aitken class D PFFD and only a remnant of distal femoral epiphysis in which knee fusion will have little to offer, this may be a suitable choice.

The Van Nes rotationplasty requires a nonconventional prosthesis with the ankle functioning as the new knee. This is a very difficult prosthesis to align and fit, although it gives excellent function (177, 194). The prosthesis has a lower padded foot socket that contains the rotated foot in full plantar flexion. Lateral and medial external joints are attached to the upper thigh section to increase stability and to prevent hyperextension of the lower shank (194). The original design incorporated a laminated thigh section with ischial weight bearing. For patients with good hip stability, for example, in those who had a tumor and trauma, the laminated section is often replaced with a leather thigh lacer and no ischial weight bearing. It is imperative for proper function that the external joints be aligned with the axis of rotation of the ankle/subtalar complex while maintaining the line of progression. Failure to ensure this alignment, regardless of the anatomic joint, will result in a poor gait pattern and skin breakdown. The prosthetist should incorporate mechanical joint placement with slight external rotation on a new prosthesis, in anticipation of the mild internal derotation inevitable during growth.

Author's Preferred Recommendations. Children with more severe forms of femoral deficiency are initially fit with an extension-type prosthesis until the age of approximately 3 years. For children with a congenitally short femur with <20 cm of anticipated limb-length discrepancy at skeletal maturity, an arc of motion of the knee of 60 degrees without flexion contracture, and a foot that is plantigrade or can be made plantigrade with surgery, the authors suggest limb lengthening. Hip and proximal femoral stability are achieved first by redirecting or augmenting the acetabulum as necessary and repairing a proximal femoral pseudarthrosis and/or varus if it exists.

For patients where the foot falls at the level of the contralateral knee, the authors suggest Van Nes rotationplasty and knee fusion if the family is accepting the idea and the ankle/subtalar joint complex has at least a 60 degrees arc of dorsi/plantar flexion and no equinus contracture. If the family is unaccepting of rotationplasty, or if the foot and ankle are not well-aligned and/or lack sufficient range of motion, then knee fusion with Boyd amputation is undertaken. Careful prediction of the ultimate length of the tibial and femoral segments guides the decision of which epiphyses and physes to remove at the time of knee fusion. In the majority of cases, both physes will be removed.

The authors do not have sufficient experience with iliofemoral arthrodesis with or without rotation to recommend either procedure.

Pearls and Pitfalls. In evaluating these patients, it is important to accurately predict the ultimate limb-length discrepancy as early as possible. This is essential in formulating an early treatment plan. Because of the flexion, abduction, and external rotation deformity of the proximal femoral segment,

traditional scanogram radiographs can overestimate the amount of shortening. Scanograms should be obtained in the lateral position, which will account for any flexion deformity at the hip (or knee) in these patients.

For patients where knee fusion and foot ablation is the treatment plan, an accurate prediction of femoral and tibial segment length at maturity will help the surgeon decide if the distal femoral and/or proximal tibial epiphysis and physis need to be removed at the same time. If one (or both) physes about the knee are removed at surgery, the physician should counsel the family that the residual limb will appear "too long" immediately after surgery, but that the normal limb will overgrow the residual one with subsequent growth.

With regard to the hip flexion, abduction, and external rotation deformity seen in these patients, the authors experience is that the deformity resolves after knee fusion and foot ablation with prosthetic use over several months. Therefore, when performing knee fusion, the tibia should be fused in line with the femur rather than in a flexed, adducted, internally rotated position to compensate for the proximal femoral segment alignment. The authors do not have experience with concurrent soft-tissue release at the time of osteotomy (so-called "super-hip" procedure).

Complications. Progressive hip subluxation and frank dislocation can occur with femoral lengthening. Prevention of this complication is the best treatment. Careful evaluation of the hip should be undertaken prior to femoral lengthening, if it is to be performed. Hip dysplasia should be addressed prior to lengthening with appropriate acetabular reorientation or augmentation procedures.

As mentioned previously, insufficient or recurrent rotation can occur after Van Nes rotationplasty. Treatment of this problem is with repeat tibial rotational osteotomy.

CONGENITAL DEFICIENCIES OF THE UPPER EXTREMITY

Definition and Classification. If upper extremity deficiencies were completely discussed like lower extremity deficiencies, the list of topics covered in this section would be much larger. Longitudinal deficiencies in the upper extremity include radial and ulnar longitudinal deficiencies, as well as symbrachydactyly and cleft-hand deformity. Thumb hypoplasia or aplasia might also be listed. In contradistinction to the lower extremity, the shortening which results from these longitudinal deficiencies is rarely the primary clinical problem as it is in the lower extremity. In reality, pediatric hand and upper extremity surgeons participate in multidisciplinary team clinics to care for these patients (much like pediatric orthopaedic surgeons participate in multidisciplinary team limb-deficiency clinics), and surgical reconstructive treatment of these patients is best described in chapters dedicated to the care of these specific disorders. Often, it is the patient with terminal deficiencies of the upper extremity that are treated in limb-deficiency clinics, are often candidates for prosthetic use, and are the focus of this section. The child with an upper extremity amputation has an inherently different disability than the child with a lower extremity amputation. The importance of sensation in the hand, which is seen as the terminal "working end" of the entire upper extremity, cannot be overemphasized. Without sensory feedback, the child with an upper extremity limb deficiency must look at the prosthetic hand to help it function. In addition, the child must think actively to control the terminal device of an upper extremity prosthesis. Both of these factors make upper extremity prostheses much less efficient and much more difficult to use than a lower extremity prosthesis. At best, any upper extremity prosthesis will be used more as an assisting limb rather than as a substitute capable of achieving bimanual function.

Additionally, upper extremity amputations are very visible. Unlike the child with a below-knee amputation who walks without a limp, has a prosthesis hidden under clothes, and can often match his or her peers in physical activity, the child with an upper extremity amputation is more easily seen as different. For both of these reasons, many patients with transverse upper extremity deficiencies will not be well served by prosthetic prescription. If the prosthesis does not afford the child a functional gain or cosmetic benefit, he or she will be quick to reject it. The reasons for some children becoming good users of a prosthesis, whereas others with the same characteristics reject it, are not well understood. Although the age at initial prosthetic fitting, as well as the parents' acceptance and compliance are important, this is not the whole answer. The incredible ability of the young child to learn to use one hand assisted by the residual limb with minimal concession to activities that are assumed to require two hands must also be a significant factor.

Quantification of successful upper extremity prosthetic use is very patient specific. The number of hours a prosthesis is used per day is not a good criterion. Many children will use the prosthesis for specific tasks (riding a bike) while preferring to remove it for others (swimming). Some children will wear and use it every day in school, but will wear it very little during the summer while playing.

What the child can do with the prosthesis when asked and what he or she actually does with it in the course of a normal day can be very different. Although some children develop an amazing facility with the prosthesis in their everyday activities, many will demonstrate their skill with the prosthesis only in the medical setting, preferring to use the prosthesis much like their residual limb during daily activities. Standardized tests have been developed to measure spontaneous use versus voluntary control as it relates to age-appropriate activities. The University of New Brunswick test of myoelectric control is used by therapists to assess the child's ability to use the prosthesis in a controlled situation. The Prosthetic Upper Extremity Functional Index is a self-reported measure of the child's functional abilities during daily activities. The Unilateral Below Elbow test is an observed functional evaluation instrument that examines both completion of specific tasks and how the prosthesis was used in that task. James et al. found that, when evaluating unilateral below-elbow amputees with and without a prosthesis,

little functional difference could be measured between the two groups when using this outcomes instrument (195).

Although children often will not use an upper extremity prosthesis for functional purposes, the importance of the cosmetic appearance of a passive hand prosthesis is of benefit to many patients. Many adolescent patients choose a cosmetic passive hand as the terminal device of their prosthesis over all other options, despite their diminished functional potential. The higher the deficiency and the more the disability, the harder it is to replace the function with a prosthesis, and the less likely the patient is to accept it. Lack of heat dissipation and functionality and the extra weight, energy expenditure, and concentration necessary to work it are all reasons for children with more proximal deficiencies to less likely use a prosthesis.

The main purpose of the upper extremities is to place the hand in space to grasp and manipulate objects. Early in infancy, the upper extremity reaches and touches objects within the visual fields, providing rich sensory feedback to the child. This feedback is an essential element of upper extremity function. For the child with an upper extremity limb deficiency, particularly a bilateral deficiency, sensation seems to be the single-most desirable attribute of the extremities. Therefore, if the residual upper extremities allow sensory feedback by meeting in the midline, the child will usually reject any type of prosthesis. If the extremities will not oppose, or sometimes, if they oppose where they cannot be seen, the patient may prefer a prosthesis for the function it affords.

The fitting of an upper extremity prosthesis is much more individualized than a lower extremity prosthesis. For those with a unilateral below-elbow amputation, fitting with a passive hand at approximately 4 to 6 months of age is an easy decision because it is relatively inexpensive and well tolerated by the patient. If the child accepts the prosthesis, this can help the health care team decide on fitting with a more complex prosthesis later. However, for higher level amputations, especially if they are bilateral, routine prosthetic fitting will frequently result in failure (196).

The age for fitting is based on the normal development of the child. By 4 months of age, the child brings the hands to the midline while supine and props on the elbows while prone. Eye–hand coordination develops as the hands are brought into the visual fields. By 6 months, the child is beginning to prop on the extended arms when sitting and is rolling in all directions. Early prosthetic fitting between 4 and 6 months allows the infant to incorporate the prosthesis in all gross and fine motor functions that are developing. Despite this common practice, it has been difficult to show that fitting at this age results in a higher incidence of adult prosthetic use or higher function. Recent studies suggest similar acceptance to prosthetic use if the patient is <2 or 3 years of age (196, 197).

Bilateral Upper Extremity Limb Deficiency. Children with bilateral high-level amputations will primarily use adaptive performance techniques with their mouth, chin, neck, shoulder, and feet. An occupational therapist is essential for promoting these techniques as well as adapting the child's environment to assist with age-appropriate activities. The benefit of prosthetic fitting for these children is controversial, because most function well by using their lower extremities for ADL. Attempts should not be made at this point to modify the child with prosthetics. Children with high levels or complete absence of the upper limbs will use their feet to accomplish everyday two-handed activities. The use of the foot in play and in ADL appropriate for the child's stage of development should be incorporated in all therapy home programs. It may take considerable persuasion to win the parents to this view.

Nonetheless, children with bilateral high-level amputations should be given an opportunity for prosthetic use. In addition to possible functional gains, the families experience a significant emotional benefit in knowing that all has been tried, and the patients gain a valuable experience in attempting to use the prostheses. A multicenter review of bilateral upper limb deficiencies showed that 50% of patients were still wearing a prosthesis at age 17 years or more (196). Fitting in such children should rarely be attempted before 1 year of age, despite the parents' anxieties. Fitting should be done to help the child perform appropriate tasks for his or her stage of development, or to aid in certain specific activities. It is usually best to fit a child with only one prosthesis at a time because the problems with two may lead to early rejection.

Specific Congenital Upper Extremity Deficiencies

Amelia. The child with unilateral absence of the entire arm will be less likely to fully accept a functional prosthesis than those with lower levels of amputation. If body-powered components are used, the patient has difficulty in controlling the devices because there is no lever arm. Externally powered prostheses are heavy. The weight and increased body heat due to the necessary suspension make this a difficult prosthesis to wear. When these facts are added to the problems of function in using an artificial shoulder, elbow joint, and hand, the child will usually choose to function without the prosthesis.

Many children with amelia of the upper extremity have bilateral deficiencies. The choices for these children are to help them develop their lower extremities to substitute for the upper extremities, to fit them with prostheses, or to attempt a combination of both. There is universal agreement that no attempts should be made to limit the child's use of the feet nor to provide all of the child's upper extremity function with prostheses. The feet are the best substitute for the hands. Children with bilateral amelia and relatively normal lower extremities can usually master all ADL, while leading full lives with family, children, and employment. This is not often understood by health care providers until they become acquainted with an older child or adult with bilateral amelia. Most of these children will reject prostheses.

The question of prosthetic fitting most often arises in the child with bilateral amelia and significant lower extremity anomalies that limit their substitution for upper extremity function. In such cases, unilateral fitting may be indicated, but is likely to gain limited acceptance, and then only after many years of struggle.

Children with bilateral amelia often walk late. They will need help in pushing to stand. In addition, the fear of falling, because they cannot protect themselves, comes early. Helmets or some protective headgear are needed until the child is independent in gait. Therapy concentrates on trunk control and strength, along with training in the use of their feet for all activities. These children are also prone to develop a progressive scoliosis, often before adolescence. This presents a difficult problem. Treatment of the scoliosis, either by bracing or by surgical fusion, restricts the use of their feet in ADL. However, often these curves will be progressive, and in such cases, a selective fusion is preferable.

Phocomelia. In phocomelia, the distal portion of the extremity appears to attach directly to the body (Fig. 30-37). Although typically described as an intercalary deficiency, a recent study suggests that the limb almost always exhibits a longitudinal deficiency proximal and distal to the intercalary defect (Goldfarb et al.). There are wide variations in the severity of this deficiency. In some, the hands may be close to normal, with some remnants of the arm bones, whereas in others the hand may be no more than a single functionless digit with near complete absence of the arm. Patients with phocomelia usually have some mobility in their residual limbs and therefore differ in one significant way from the child with bilateral amelia—they have a sensate limb often capable of some grasping function. The function of these limbs depends on the function of the hand, the length and function of the arm itself, and the ability to bring the hands together within the field of vision.

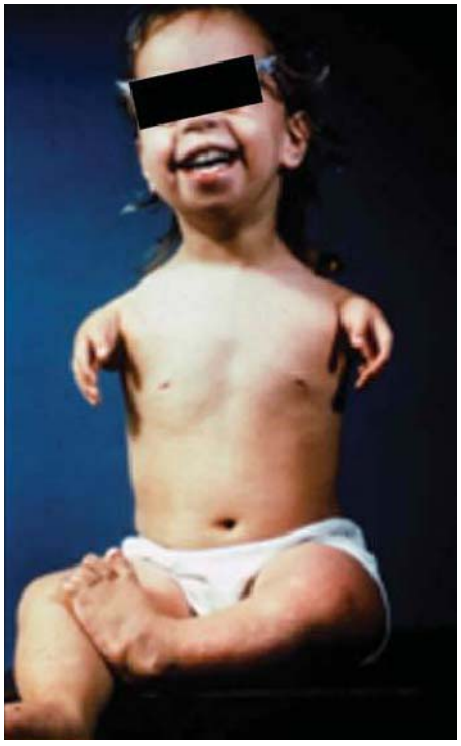


FIGURE 30-37. Clinical photograph of a patient with bilateral upper extremity phocomelia, with the hands seemingly arising directly at the level of the shoulder joint. Patients with phocomelia often have the ability to use their residual limbs, but in cases like this they rely on their lower extremities for ADLs.

As mentioned previously, those children who cannot bring their hands to the midline or to the mouth will use their feet to substitute in the ADL (Fig. 30-38A–D). They should be encouraged from an early age to develop their foot skills and the body strength that is necessary to use the feet. In those children with longer residual limbs and better function, little treatment other than providing adaptive equipment to aid in dressing and so on may be necessary (Fig. 30-38E).

Therapy to increase the range of motion of the scapula and limbs and to strengthen any muscle power in the residual digits may be beneficial. Adaptive equipment can be very useful for some activities such as feeding, dressing, and so on. The residual limbs can manipulate switches for powered prostheses, giving rise to the temptation to find a prosthetic solution to their problem. However, like the child with bilateral amelia, these children will function in most activities by substituting foot function for what they cannot do with their upper extremities. Fitting the older child with a unilateral prosthesis for a specific function may be indicated, but routine wear is not common.

Transverse Complete Humeral Deficiency. There is little published experience in this deficiency. Although some children with unilateral above-elbow amputation will develop surprising facility with a prosthesis, most will often wear it only for specific activities, such as sports, or for cosmetic reasons in social situations. In children with a long-enough humeral segment, the humeral–thoracic pinch provides useful assistance for the normal opposite extremity. Prosthetic use in these children generally relates to specific tasks. These may be as limited as for riding a bicycle or full-time use at school. An attempt at prosthetic fitting is warranted.

Patients with bilateral congenital transhumeral deficiencies are more inclined to use their own body, rather than a prosthesis. These children may benefit more from assistive devices than from prostheses. Such patients will often prefer prosthetic fitting on one side, where they use the humeral–thoracic pinch, with intact sensory feedback on the other side and in their feet.

For the patient with bilateral above-elbow transhumeral deficiencies, Marquardt has recommended an angulation osteotomy of the humerus (197). The osteotomy angles the distal 3 to 5 cm of the humerus anteriorly by 70 to 90 degrees. This allows for suspension of the prosthesis without the usual shoulder cap, makes it easier to put on, permits better shoulder motion, and gives better control of rotation. This procedure should only be performed if the humeral length is sufficient and there is a need for a unilateral prosthesis.

Transverse Complete Forearm Deficiency (Congenital Below-elbow Amputation). The congenital below-elbow amputation is the most common of all of the upper extremity deficiencies. It is more often the left arm (Fig. 30-39). It is sporadic and without known cause.

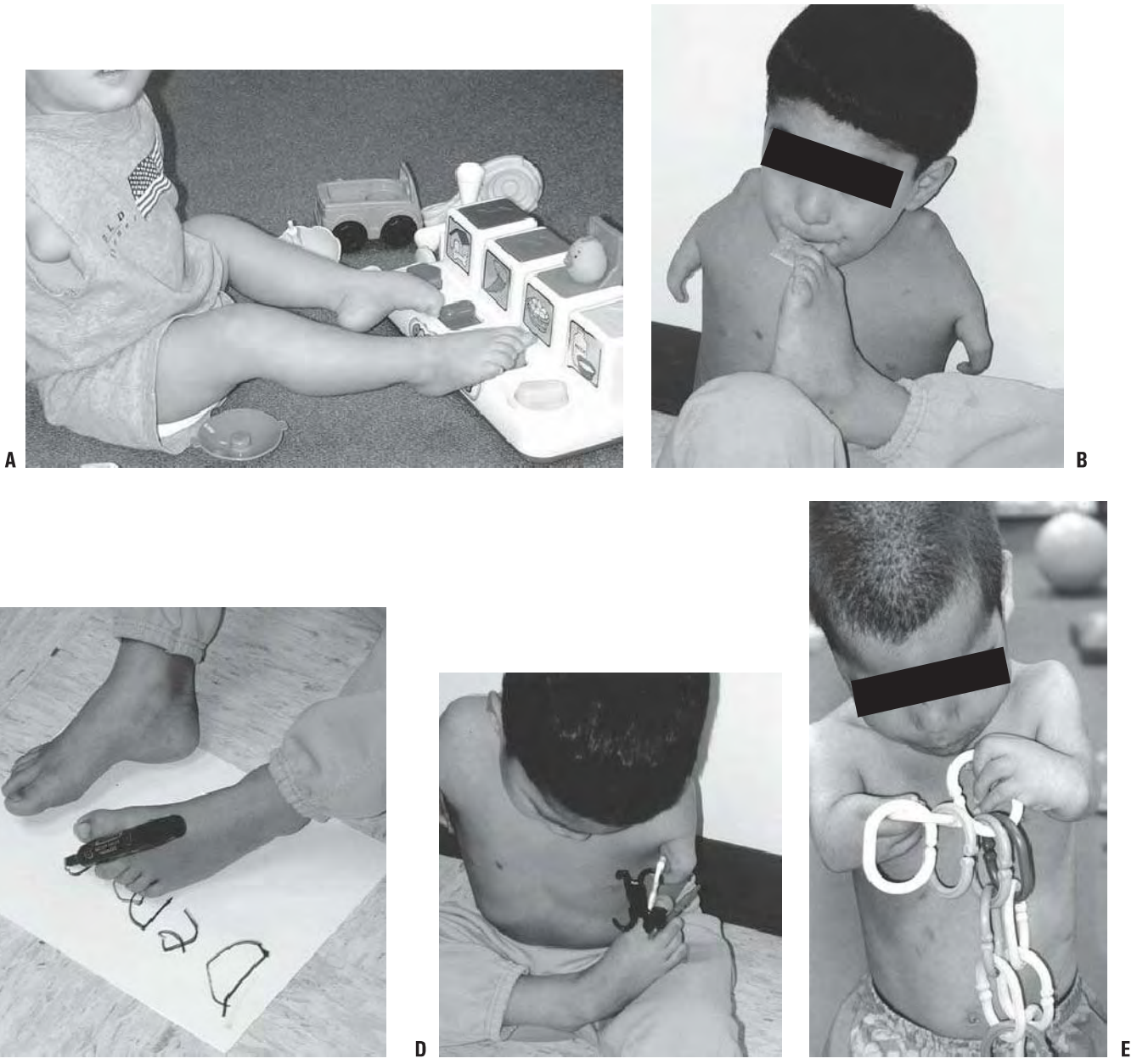


FIGURE 30-38. A child with bilateral amelia or bilateral phocomelia, when the hands cannot meet in the midline, will use the feet for most activities. In such children, the use of the feet should be encouraged and developed from an early age **(A)**. When older, these children will need the use of their feet to accomplish the activities of daily living **(B,C)**. If there is any motor power in the extremities, they may be capable of useful function and assist the feet **(D)**. When the hands can meet in the midline and have good motor power, excellent function is possible **(E)**.

Functionally, these children have a more favorable natural history than one might initially surmise. Quality of life and functional outcomes in these patients demonstrate good function as compared to normal peers (195). Nevertheless, children with this deficiency present the most ideal upper extremity deficiencies for prosthetic fitting. Children with this deficiency are unlike those with a transverse amputation through the carpal bones, which usually have partial grasping function with sensation. They are also unlike the above-elbow deficiencies in that they have a normal shoulder and elbow that allow accurate placement of a relatively light prosthetic terminal device.

Despite being ideal candidates for prosthetic use, not all children with congenital transverse forearm deficiency will remain good prosthetic users during their childhood. Scotland and Galway reviewed the experience at the Ontario Crippled Children's Center and found that 32% had stopped using their prosthesis, upon follow-up of 7 to 17 years (198). How many of these may resume use of the prosthesis while employed is unknown. They, like Brooks and Shaperman (199), noted greater acceptance of the prosthesis if fitting was done before the age of 2 years.

In the congenital group of Brooks and Shaperman (199), 22% of those fitted before 2 years of age, and 58% of those

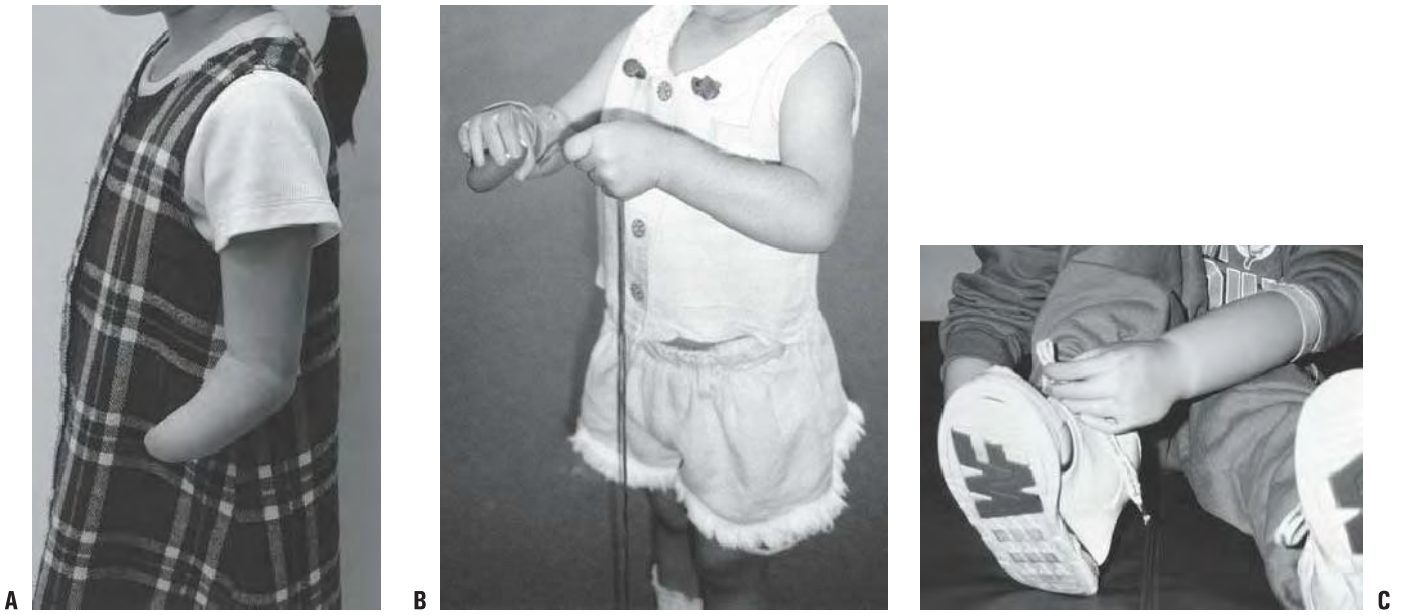


FIGURE 30-39. **A**: A typical patient with a congenital below-elbow amputation. There is usually enough length to fit a prosthetic arm and still permit good active elbow motion. **B,C**: Two different children with transverse incomplete forearm deficiency fitted with a myoelectric-powered hand performing common functions of childhood. Many of the children who use the prosthesis develop amazing skills in its use.

fitted after, had stopped using their prosthesis. However, of those who continued to use their prosthesis, there was no difference in the amount of use between the two groups. The most common age at which patients discontinued use of their prosthesis was at 13 years, most commonly because the prosthesis was viewed as cosmetically unacceptable and functionally superfluous. Sorbye (200) reported that, of the patients in their clinic who were younger than 24 years, 87% were using their myoelectric prosthesis, and 65% of these used it all day and for all activities. Hubbard et al. (201), reporting on the Toronto experience, found that 70% of the below-elbow amputees were using their prosthesis, whereas 30% had rejected it.

With this and other evidence, it is now the usual practice to recommend fitting around the age of 4 to 6 months with a passive hand to aid in normal development. This lightweight prosthesis helps the child become comfortable with a prosthesis and acquaints them with the two-handed activities that a normal child would perform. The hope is that the child will develop the central cortical pathways necessary for bimanual dexterity.

Depending on the child's acceptance and use of this passive prosthesis, a more functional terminal device is fit between 15 and 18 months (31, 202). Today, there are a number of terminal devices available (203). There are two choices to power the device: battery (myoelectric) and body (cables). Although there will be many factors to consider in the selection (cost and funding, clinic philosophy, and parent choice), virtually all centers today in North America are fitting most children with myoelectric powered terminal devices (Fig. 30-39). Table 30.4 compares the advantages and disadvantages of myoelectric and body-powered terminal devices for the child with a congenital below-elbow amputation.

Surgical Treatment Recommendations. One surgical option in the treatment of patients with bilateral transverse forearm deficiency is the Krukenberg operation. This operation separates the radius and ulna to create a forearm capable of pinch and grasp between sensate ends (Fig. 30-40). This was invented in 1916 to treat World War I upper extremity traumatic limb deficiency patients, and there are several favorable reports of function after this surgery for traumatic amputation in the literature (204–207). The procedure has also been proposed for patients with congenital transverse forearm amputation with similar good results (208, 209). This procedure has been accepted in third-world countries for both congenital and traumatic bilateral upper extremity amputations. In the Western world, concerns over the cosmetic appearance of the arm after surgery have limited its use, much like the Van Nes rotationplasty. Current surgical indications are limited to the blind bilateral upper extremity limb deficiency patient.

The advantages of the Krukenberg procedure are that the child gains sensory feedback with pincer function between the distal radius and ulna, which cannot occur with a prosthesis. Moreover, the operation does not preclude prosthetic fitting. At the author's institution, the experience with the Krukenberg procedure is that patient's Krukenberg limb becomes the dominant functioning extremity. Patients occasionally will choose to wear a passive hand cosmetic prosthesis in certain social situations over their Krukenberg limb. Complications including

TABLE 30.4 Comparison of Myoelectric with Body-powered Prostheses

Myoelectric	Body-powered
Weight Heavier; passive prostheses are weighted to prepare the patient for the myoelectric system	Weight spread across the shoulders
Grip Strength Stronger than a body-powered prosthesis; strength built into the system; less work for the child	Strength is provided by rubber bands that increase or decrease the tension and force needed to open the hand; more work needed through scapular control
Cosmesis More accepted; resembles the hand; more self-esteem for the child; greater parental acceptance	Hooklike or clawlike in appearance; usually rejected by the child or parent; object of ridicule by other children
Maintenance Requires gloves; requires maintenance and electronics; electrodes lose contact and require occasional adjustment; adjustments required for growth	Requires gloves if mechanical hand is used; frequently requires repair for broken cables caused by friction; requires adjustments for growth on a regular basis
Harness No harness or straps on body; suspension through socket	Harness is bulky on a toddler, difficult to keep on without pinning to undergarment; children dislike harness especially in summer months
Muscle Control Uses muscles in limb to control terminal device; with prosthetic hand, the child can experience direct "cause and effect"	Uses muscles distant from the terminal device, biscapular abduction to control hand or hook; very confusing for toddler whose control and balance is developing; easier for older children
Grip Control Maintains grip more easily through muscle contraction at the end of the residual limb; is the most successful type of control	Toddlers lose grip very easily, overall gross movements allow the object being grasped to release and fall involuntarily; tension on the cable to the harness does not remain constant; becomes very frustrating for the child

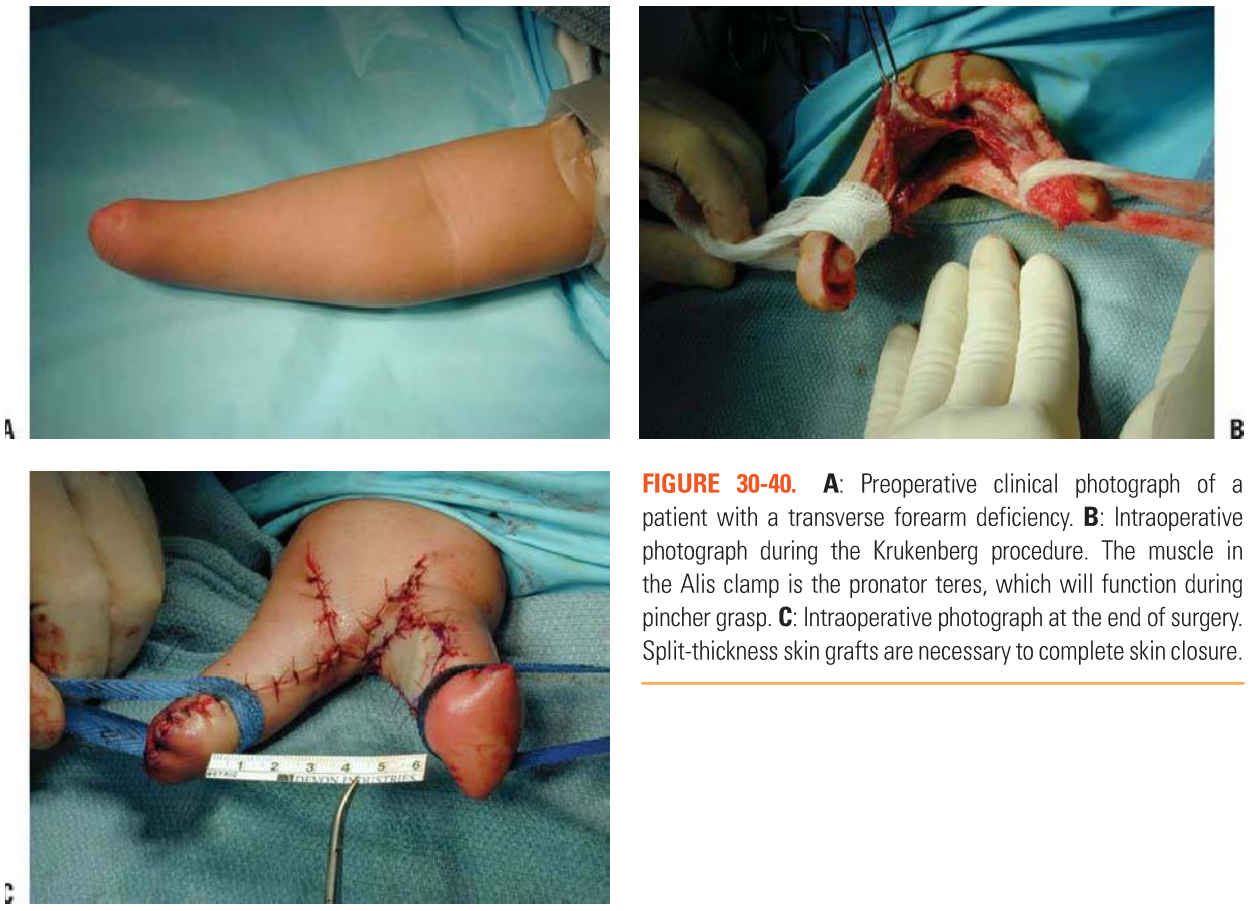


FIGURE 30-40. **A**: Preoperative clinical photograph of a patient with a transverse forearm deficiency. **B**: Intraoperative photograph during the Krukenberg procedure. The muscle in the Alis clamp is the pronator teres, which will function during pincher grasp. **C**: Intraoperative photograph at the end of surgery. Split-thickness skin grafts are necessary to complete skin closure.

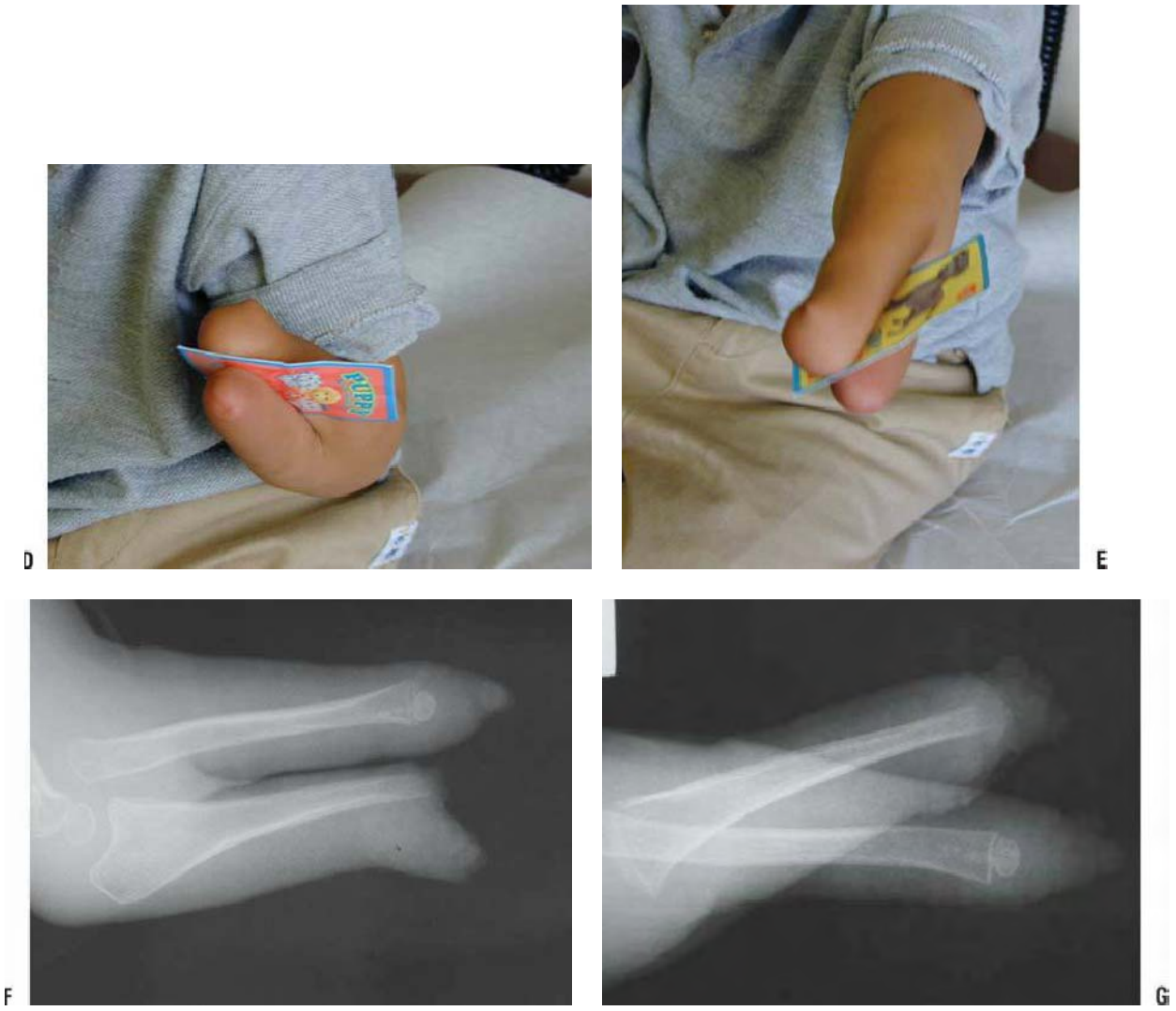


FIGURE 30-40. *(Continued)* **D,E**: Postoperative photographs of the patient demonstrating pincer grasp of a playing card. **F,G**: Forearm radiographs of the same patient taken in supination and pronation. The patient opposes the tips of the residual limb with pronation. Note the dislocated radial head seen in supination. Occasionally, this can become symptomatic.

proximal radial head subluxation or dislocation can be a problem. Also, a minority of patients lack sufficient pincer power to take full advantage of the procedure's functional benefits. The authors recommend this procedure in a child with bilateral upper extremity limb deficiency on one side if there is full elbow and forearm motion and the forearm is long enough to have a properly functioning pronator teres insertion (25% of the normal forearm). Family counseling before surgery is essential, which includes showing a video of patients before and after the operation and/or meeting a patient who has had the procedure.

Another surgical option for the short transverse forearm amputation is residual limb lengthening (56, 210, 211). Bernstein et al. reported that this procedure could convert a child with a short below-elbow amputation that was incapable of being fit with a below-elbow prosthesis to a standard below-elbow prosthesis. The procedure is not without complications, including elbow subluxation or contracture, and soft-tissue coverage difficulties at the distal residual limb end. The authors recommend this procedure when the residual limb is incapable of being fit with an appropriate prosthesis for the level of amputation.

Terminal Transverse Transcarpal Deficiency. The congenital transcarpal amputation is the second-most common deficiency of the upper limb and occurs in a characteristic pattern, with varying degrees of preservation of the proximal carpal row. The existing flexion of the carpals on the radius allows for limited grasping function, which along with normal sensation, makes this an assistive hand for which no prosthesis can substitute (Fig. 30-41). Occasionally, children will benefit from a volar opposition post for certain activities. They will usually wear it only for certain tasks, for example, as a guitar pick adapter or to grasp the handle bars on a bicycle.

The authors' experience with such children is that they have much more difficulty with the cosmetic aspect of their deficiency than do those children with transverse amputations

FIGURE 30-41. This young boy with a transverse transcarpal deficiency demonstrates the partial grasp at the flexor crease that, when combined with sensation, usually proves superior in function to a prosthesis.

at higher levels. Some older children and adolescents will desire a cosmetic hand that would be used in certain circumstances or would provide a psychosocial benefit. Some cosmetic hands have a passive spring grasp to provide limited function.

Prosthetic Management. Generally, upper extremity prostheses and their control systems can be subdivided into three categories: *passive, externally powered*, or *body-powered devices*. The Ballif arm (circa 1400) was the first body-powered prosthesis to introduce the use of prosthetic hand operation by transferring shoulder movement to activate the terminal device (212). A harness over the contralateral shoulder is connected with a thin cable and housing to a terminal device. Through scapular abduction, the fixed cable is stretched over a greater distance and causes the prosthetic hook or hand to open or close, depending on the configuration of the terminal device. A good analogy is using a hand lever to activate the brakes on most bicycles. Most parents prefer a prosthetic hand over the cantered hook for cosmetic reasons. Unfortunately, the hook is far superior in function, but has fallen from favor because of the desire to have the prosthesis look as natural as possible, even at the sacrifice of function. Most hooks are canted in design, and this allows the child to see what is being grasped, as compared to the mechanical hand that obstructs the view and results in awkward arm positions to grasp objects.

The externally powered prostheses are powered with motors and can be further subdivided into *switch control* or *myoelectric control*. In both systems, a battery, relay switch, electric hand, and electronic control system are present. It should be noted that the myoelectric hand is the only terminal device available for children using the externally powered prosthesis. In the myoelectric prosthesis (Fig. 30-42), the child contracts various muscles in the residual limb voluntarily, and an electrode placed on the surface of the skin acts to pick up the electromyographic signal. The signal is in turn amplified with the help of an electronic relay switch, and this, in turn, operates the electric hand (213). The entire system is generally referred to as a one- or two-site system. This denotes the number of electrodes that are used for signal recognition.

The one-site system can be further categorized as voluntary opening–automatic closing, rate sensitive, and level sensitive. Myoelectric arms are generally fit before age 2 and utilize a voluntary opening–automatic closing (cookie-cruncher) configuration. Muscle contraction opens the electric hand, and relaxation causes the hand to close automatically. After age 4 years, more advanced prostheses can be fit. These include rate-sensitive and level-sensitive control systems, which use one muscle to control two functions, and are generally fitted to children over 4 years of age, when two sites are not available. Because muscle contraction controls more than one joint in this case, the prosthesis is more difficult to learn to use. The choice of a system depends on the muscle signal strength, muscle control, and prosthetic design factors (31).

In the two-site system, each electrode operates a specific task. Children can often operate this more complex system by 3 to 3 1/2 years of age. Contraction of wrist flexors closes the hand, whereas contraction of wrist extensors is used to open the hand. This system is used when children have demonstrated good control and use of their myoelectric prosthesis and can control both the flexors and extensors independently of each other.

Patients with a higher level of upper extremity amputation are generally good candidates for switch-controlled externally powered prostheses. The electrode is replaced with a miniature switch that can be of a push–pull configuration, a force-sensing resistor, or of a simple toggle design. The incorporation of these switches into the prosthesis depends primarily on the level of amputation and the design of the prosthetic socket or frame.

Multiple Limb Deficiencies. Management of the patient with multiple upper and lower limb deficiencies is a challenge that requires a team with experience to achieve the maximum function for the patient. The difficulties of bilateral upper extremity amputation have been covered earlier. Children with one upper and one lower extremity pose no

FIGURE 30-42. This example of a myoelectric prosthesis, called the Otto Bock Electrohand, was made with a clear socket for teaching purposes. The proximal portion of the socket, which fits on the residual limb, contains the electrodes that pick up the signals from the muscles. This fits into the prosthesis, which contains the electrical and mechanical working parts of the hand.

problems beyond the management of each individual limb. Children with bilateral knee disarticulation or transtibial amputations will walk without support, and therefore a unilateral upper extremity amputation in association poses no special problem, other than donning and doffing the prostheses. With bilateral amputations above the knee disarticulation level, however, walking without support is problematic; upper extremity function is needed, and a wheelchair may be required for long distances and to conserve energy.

One of the most common types of patients seen in the pediatric age-group with this problem is the quadrimembral amputee resulting from neonatal meningococcemia (Fig. 30-43). In these patients, it is often necessary to cover the residual extremities with split-thickness skin grafts to maintain length. These grafts, if not adherent to bone, hold up very well in the prosthesis, and are not a hindrance to fitting. Treatment must be individualized for each patient, while following certain general guidelines. The first is to help the patient maximize function with his or her residual limbs. This is especially true with the upper extremities, in which sensation is so important to function. Although these children will become proficient in the use of bilateral upper extremity prostheses, if their residual limbs are long enough, they will usually perform many of their daily activities, especially at home, without their prostheses.

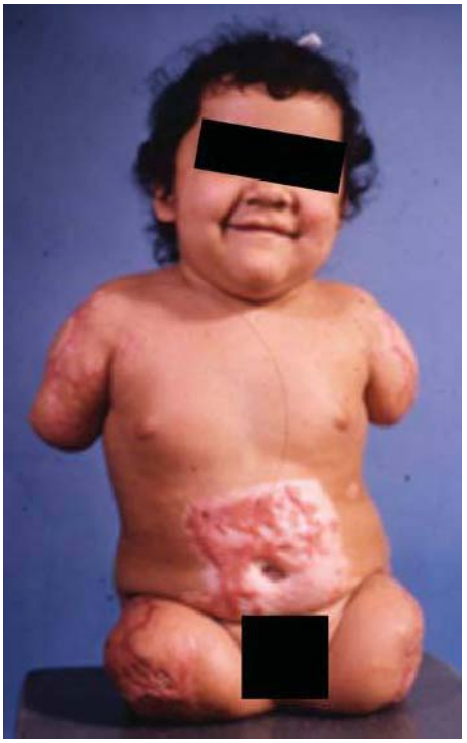


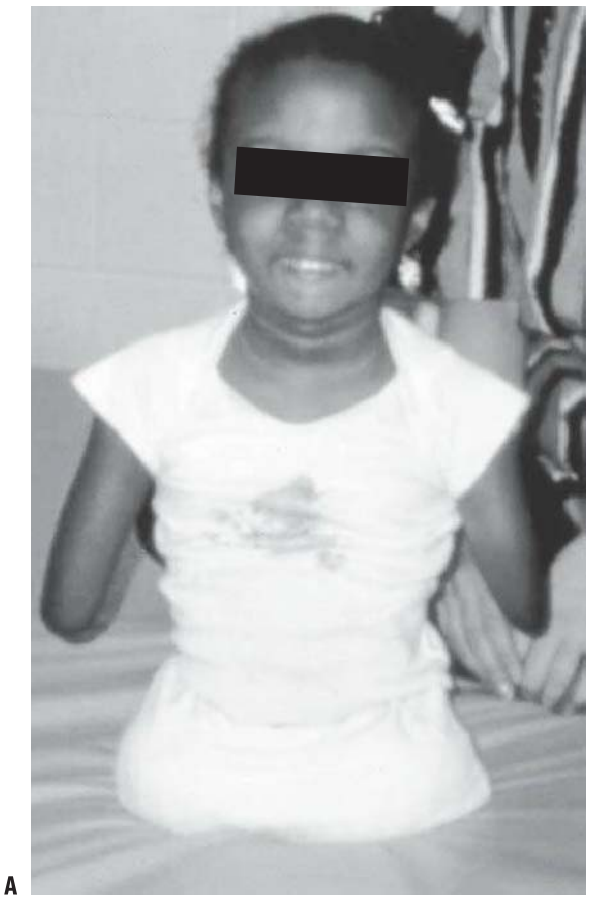
FIGURE 30-43. Clinical photograph of a patient with meningococcemia and quadrimelic limb deficiency. Multiple split-thickness skin grafts were necessary to cover the residual limbs after infection.

A common mistake is to attempt to fit all four extremities of these children with quadrimembral loss at the same time. Doing so may result in actual delay in functional recovery and rejection of the prostheses. In most situations, it is best to first fit the lower extremities and achieve ambulation, then fit the upper extremities.

In the child with a congenital quadrimembral deficiency, there will usually come a time when the parents, and perhaps the child, desire prosthetic fitting. As is true for acquired deficiency, it is best to avoid fitting all four extremities at once, but rather focus on meeting specific needs. Although experience shows that most of these children will have limited or no use of their prosthetic devices, they and their parents need and are entitled to this experience at least once (Fig. 30-44).

Authors Preferred Recommendations. The authors generally fit patients with upper extremity prosthesis with a passive hand terminal device when sitting balance is achieved. The child is fit with an activated terminal device between 1.5 and 2 years of age. No generalizations as to the type of terminal device can be made. As mentioned previously, multiple limb-deficient patients are fit with lower extremity prostheses first, followed by upper extremity prostheses within the first few years of life. The fitting of the child with upper extremity deficiencies is highly patient dependent.

Patients with bilateral transverse forearm amputations with residual limbs of sufficient length are offered a unilateral Krukenberg operation at school age. Patients with unilateral transverse forearm deficiency with a short residual limb unable to be fit with an appropriate level prosthesis are offered forearm lengthening to improve prosthetic fit and function.



A

B

C

FIGURE 30-44. This child with congenital bilateral hip disarticulation and transhumeral amputations **(A)** was fitted with four prostheses **(B)**, which she quickly abandoned in favor of her power chair and simple assistive devices **(C)**.

Pearls and Pitfalls. Children with bilateral amelia will depend on their lower extremities for all of their ADL. It is important to refrain from any lower extremity surgery until the full extent of lower extremity use is understood. It is especially important to avoid any lower extremity amputation in these children if it interferes with their ADL. Also, special considerations for treating spinal deformity in these patients are warranted. Because spinal flexibility may assist in allowing the lower extremities to substitute for the upper extremities, preservation of spinal motion is a special concern. If spinal fusion is necessary, limited fusion is indicated.

Patients with transcarpal amputations often have radiocarpal motion with limited pinch function. It is difficult to improve function in these children, either with prosthetic fitting or with surgery.

In the child with four-extremity limb deficiency, it is best to hold off on fitting upper extremity prostheses until the child has been fit with lower extremity prostheses and begun walking.

Complications. Complications of the Krukenberg operation have been previously mentioned and include radial head subluxation progressing to dislocation as well as inadequate pincer power. With regard to lengthening residual transverse forearm residual limbs, elbow subluxation and/or contracture are possible complications, along with the usual complications that go along with limb lengthening of any sort.

ACQUIRED DEFICIENCIES

Causes. Children may undergo an amputation for a variety of reasons. Although there are no good statistics, trauma is the major cause of amputation in childhood (214). In this group, power lawnmowers lead the list of causes, and most commonly, it is a young child riding on the lap of a family member. Motor vehicle accidents (particularly all-terrain vehicles), farm injuries, and gunshot wounds follow in that order (215). In war-torn countries, landmines may be the leading cause of amputation. Because amputation of the digits is most often caused by machinery, upper extremity amputations are more common than those of the lower extremity. Boys are affected about twice as often as girls.

Tumors, vascular occlusion caused by meningitis or vascular catheterization, and burns are additional causes, and each has its own unique set of circumstances. Indeed, the differences in acquired amputation defy classification. Some are semielective and allow for some preparation of the patient and the parents, whereas traumatic amputations do not. It is possible, however, to discuss the general principles that are applicable to acquired amputations in children.

Principles. When the surgeon is faced with an acutely mangled extremity, it can be difficult to decide on amputation versus limb salvage. Treatment principles in adults are not easily transferred to children. Most often, multiple physicians should discuss the case to make the best assessment, while always remembering the tremendous healing and adaptive capacity of the child, compared with the adult.

When dealing with lawnmower injuries to the foot, it is often wise to attempt limb salvage at the first surgery. However, for more proximal lawnmower injuries that will require vascular and nerve repair, along with bone reconstruction and free tissue transfer, the decision needs to be made more realistically. The more energy expended in saving the limb, the higher the parents' expectations of the result.

The surgeon will rarely have options regarding the level of amputation in trauma. This usually dictates saving as much of the limb as possible. This often results in a less functional level of amputation than one that is more proximal. The most obvious example is preservation of the talus and calcaneus, which the parents and child may see as saving at least part of the foot, whereas the result may be a worse gait, worse cosmesis, and more prosthetic problems than with a Syme amputation.

In the child, it is usually advisable to preserve as much length as possible in amputations of the long bones. This is especially true in the femur, in which 70% of the growth of the bone occurs from the distal physis. A 5-year-old child with a midthigh amputation will have less than ideal length as an adult. In the lower leg, however, little is lost, so long as an adequate portion of proximal tibia, in which most of the growth occurs, is preserved, and in fact, shortening the bone to achieve good soft-tissue coverage may be the best course.

The adult dictum, that skin grafts do not make suitable coverage for a residual limb that will bear weight in a prosthesis, is not applicable to children, especially very young ones. In children, skin grafts do make good coverage as long as they are not adherent to the bone. Split-thickness skin grafts are frequently needed to preserve length in meningococcemia, burns, and some cases of trauma. In older children with traumatic amputations, free vascularized flaps can provide excellent coverage.

Where possible, disarticulations are preferred over through-bone amputations because they will prevent the problem of bony overgrowth. It is not necessary, or perhaps even advisable, to remove the cartilage from the bone end. Tapering of the bone ends, at the distal tibia, for example, is not necessary unless the child is approaching adulthood. The bony prominences will not develop to adult proportions and therefore do not present a prosthetic fitting problem. If a young child has a through-bone amputation, it may be possible to salvage a portion of bone and cartilage from the amputated part for capping the bone. This is similar to performing a Marquardt procedure and has the potential to substantially reduce problems of overgrowth.

It is important not to forget the child during the acute phase of the amputation. Often the surgeon expends a great amount of energy dealing with the parents' emotions and can easily forget that the child also needs emotional as well as physical attention.

In many circumstances, the amputation will be elective. Such is the case with children who have posttraumatic injuries and are electing surgical modification for better function and prosthetic fitting. Children with neurofibromatosis, Klippel-Trenaunay-Weber syndrome, and malignant tumors not suitable for limb salvage also are in this category. In many cases, the need is obvious, and the child and parents have accepted their decision after careful consideration.

In the case of tumors, however, it is usually not so easy, and generally there is not complete acceptance of what is in fact a life-saving procedure. In all cases, the more preparation by the medical professionals and opportunities for the parents and patient to talk and see other patients, the better. It needs to be emphasized that the challenge to be overcome with treatment is to live, and that the surgery is necessary for that. The options revolve around the functional and cosmetic aspects of the different procedures.

There remains a difference of opinion about the benefit in fitting the acquired juvenile amputee in the immediate postoperative period. In the young child with a congenital deficiency, there seems little to be gained. However, in the older child, especially when the amputation is caused by trauma, there can be large psychological benefits from placing the child immediately in a postoperative prosthesis. This also aids in edema control diminution of phantom pain.

PROSTHETICS

In the prosthetic fitting of the pediatric amputee, the single-most important guiding principle is that functional concerns always override cosmetic ones. When dealing with the adult population, overall biomechanical forces resulting from prosthetic alignment can do relatively little damage to skeletal integrity. This is not the case for the pediatric patient, in whom skeletal development is ongoing. Incorrect alignment can have long-term and often pronounced negative results.

Role of the Prosthetist. The role of the prosthetist is to ensure that the highest level of functional need of the patient is met through prosthetic intervention, or through no intervention at all if indicated. The skilled prosthetist can assess anatomic and functional deficiencies and recommend socket design and component selection. In recent years, there has been a tremendous increase in the prosthetic innovations and components available for the pediatric amputee. Knowledge of these components and their appropriate use will generally be the responsibility of a prosthetist with special interest and experience with children. In addition, he or she must possess the clinical skills, medical knowledge, and communication skills to timely direct the flow of knowledge to the other team members and parents, so that realistic expectations can be identified and achieved. Routine maintenance of the prosthesis is extremely important so that extensive repairs will be minimized and the need for a new prosthesis recognized. Children are generally not happy to be without their prosthesis.

Fitting Techniques. The technique will vary, on the basis of prosthetist experience, team philosophy, integration of ever-changing technology, and severity of the deficiency. Physiologically, the child is in a constant state of growth and the prosthetic device must be designed both to permit weight bearing and to allow for the greatest amount of growth without compromising fit and function. Most congenital lower extremity amputees are able to bear some weight on the distal end of the residual limb, allowing the prosthetist to achieve a slightly less intimate fit of the prosthetic socket than might be the case for the acquired adult amputee. Unlike the adult, the child's skin has greater tolerance to skin breakdown. The increased activity levels of the child amputee place tremendous expectations on the prosthetic devices and the components. All of these factors are constant challenges to the prosthetic prescription and emphasize the need for a fluid approach to fitting. What may be suitable for one child may be unsuitable for a different child with the same anomaly. Rigid time schedules are discouraged, and developmental levels should be used only as a rough guideline to aid the practitioner.

Fitting the child with limb deficiency leads to unique issues not normally seen in the adult population. Knee disarticulations can lead to long residual limbs with knee centers lower than on the sound side. Ischial containment sockets are not normally recommended (unless needed because of hip instability or non-distal weight bearing) because of problems with soft-tissue containment and diapers in infants. Location of bony landmarks is obscured by fatty tissue, and casting is difficult and nonexact.

The various stages involved prior to fitting a prosthesis are generally standard within the profession. Upon referral to a clinic, the child is assessed by the team, and a treatment protocol is established. The stages involved in prosthetic fitting include:

1. *CASTING* of the residual limb.
2. *TEST FITTING* of the modified interfacing socket.
3. *DYNAMIC ALIGNMENT AND GAIT TRAINING.*
4. *DELIVERY* of the completed prosthesis.

Socket Design and Suspension Systems. The cast or impression forms the foundation for the prosthetic design (216). It is only after a well-fitting and comfortable socket–skin interface is achieved that the additional components can be added and expected to function as designed. Casting usually involves the placing of a casting sock on the residual limb, marking all landmarks and wrapping circumferentially with plaster or synthetic bandage. This becomes the negative impression. It is then filled with molding plaster and stripped, forming the positive cast ready for modification. This positive cast is then modified to distribute forces and relieve pressure in the socket for proper hydrostatic control of the residual limb.

Through the use of a clear test or diagnostic socket fabricated over the positive mold, the practitioner is able to ascertain the areas of high and low pressure and to ensure that they are directed over the appropriate areas. Common fitting problems can be flagged and corrected before the final socket design.

Computer-aided design/computer-aided manufacture (CAD/CAM) has been used as an alternative tool to plaster casting and modification of the prosthetic socket. In its most simplified form, a residual limb is scanned with an optical laser. The information is relayed to a computer, with which modifications can be made to the scanned shape to allow for increased or decreased weight-bearing areas. The finished design is transferred to a computerized milling machine to form a positive model. This, in turn, is used to fabricate the finished device. Slowly, CAD/CAM is becoming more widely used within the profession, because of advantages of design reproducibility, record keeping, and flexibility in remote locations (217). Its advantages in the pediatric setting have yet to be proven. All current CAD/CAM systems rely on surface topography of the residual limb and therefore disregard crucial data such as tissue density, tissue mobility, and underlying skeletal structures (218).

During dynamic alignment in the crawling infant, the prosthetist initially focuses on creating a prosthesis that will

aid the infant in preparation for ambulation, transitioning from crawling to standing, with little consideration of gait at this point. Sutherland concluded that mature gait patterns were established by 3 years of age (219), whereas others place the time frame closer to 6 years of age. Early infant gait patterns are, in fact, the processes of suppression of primitive reflexes and the acquisition of postural responses (220). Dynamic alignment is the manipulation of relative position of the socket to the foot and knee while the prosthesis is moving through the various stages of gait. Through the use of alignment mechanisms in the components, the prosthetist is able to shift, tilt, and rotate the knee and foot in relation to the socket. Once independent gait is established in the infant, refinement to gait can be achieved through further prosthetic alignment.

Lower Extremity Socket Design. The design and fitting of lower extremity prostheses encompasses numerous biomechanical principles and their application to the residual limb–socket interface, and it is the successful manipulation of these forces that ensures a patient's comfort and function. The accommodation for differences in tissue compressibility, pressure tolerance, underlying bony structures, and vascular integrity are factors taken into account prior to socket design. Dynamic forces exerted through ground reaction forces and resulting moments, including torque and shear forces, increase the vulnerability of the skin–socket interface.

Hip Disarticulation. Amputees at the hip disarticulation level require extensive prosthetic intervention. The socket encompasses the amputated pelvic remnant and encloses the contralateral side for suspension. The traditional socket design rises proximally to the waist and fits similarly to a Boston spinal orthosis. The diagonal socket is a modified version of the standard design, and it affords a more comfortable fit and increased flexibility. Prosthetic hips arc on a single axis and are mounted on the outside anterior distal aspect of the affected side. Hip and knee flexion are easy to activate through pelvic tilt, if the prosthesis is perfectly aligned. The hip is anterior to the weight line and the knee is posterior. This allows for a stable stance and a smooth gait. The endoskeletal system is used exclusively for this level of amputation because of an increased level of cosmesis and decreased weight. Children with amputations at this level can achieve remarkable gains when fitting begins while the child is pulling to stand and when therapeutic intervention and parental training are incorporated. It is recommended that the knee be locked, initially, so that hip control can first be learned. Once the child is walking independently, the knee can be activated.

Transfemoral Prosthesis. Transfemoral socket design has evolved significantly over the last 10 years. The quadrilateral socket was the socket design of choice until 1987, when the ISPO formulated recommendations on the narrow medial lateral (ML) socket design (221). Variants of this design (ischial containment socket) continue, including the contoured adducted trochanteric-controlled alignment method (CAT-CAM), the normal shape, normal alignment, and the modified quad designs, to name a few (Fig. 30-45). The underlying principle is to adduct the femur while locking the ischial tuberosity within the socket, thereby providing a more anatomically correct alignment during all phases of gait (222). The controversy over these designs has been increasingly dispelled, with further clinical experience.

There are various suspension mechanisms that may be utilized for the secure attachment of the socket to the residual limb. These devices may provide auxiliary suspension which is attached to the socket to suspend or enhance suspension. The suspension may be incorporated in the socket itself, as in suction sockets, SC sockets, and so on.

Pediatric amputees are usually fitted with a *Silesian belt* system of suspension (Fig. 30-46A,B), until adequate development of the residual limb allows for silicone suspension, at approximately 2 to 3 years of age. The Silesian belt attaches to the anterior/medial aspect and the lateral aspect of the transfemoral socket and lies across the pelvis at the waist. Tightening the Silesian belt prevents the socket from slipping distally. The TES belt may be used instead of the Silesian belt. The TES belt is a neoprene suspension system that is applied over the proximal portion of the transfemoral socket and is then secured around the waist, and it has become the suspension of choice for the first-time prosthetic user.

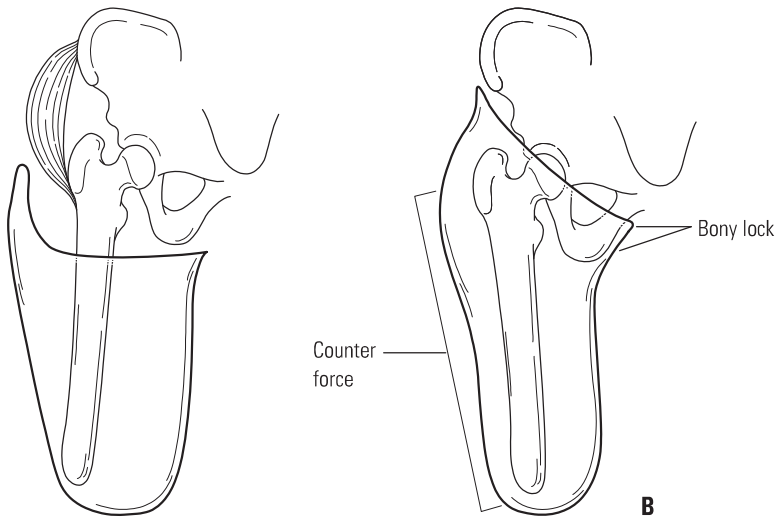
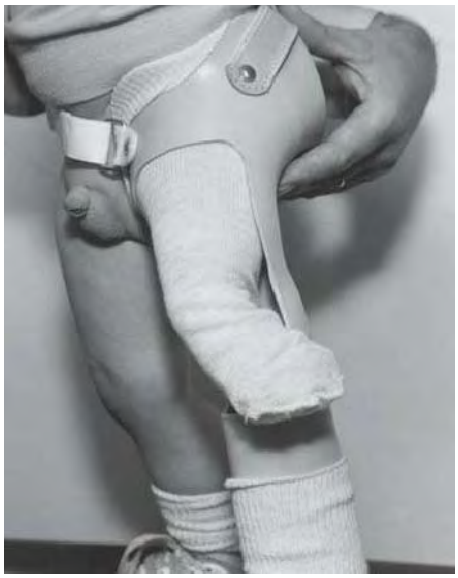


FIGURE 30-45. **A**: The quadrilateral socket is useful for the young child, especially if end bearing is possible. However, it fails to stabilize the femoral segment in a transfemoral amputation. **B**: This has led to the popularity of the narrow ML socket design, such as the ischial containment socket shown here. This design can prove impossible in small infants, because of the fatty thigh and buttocks as well as the diapers.



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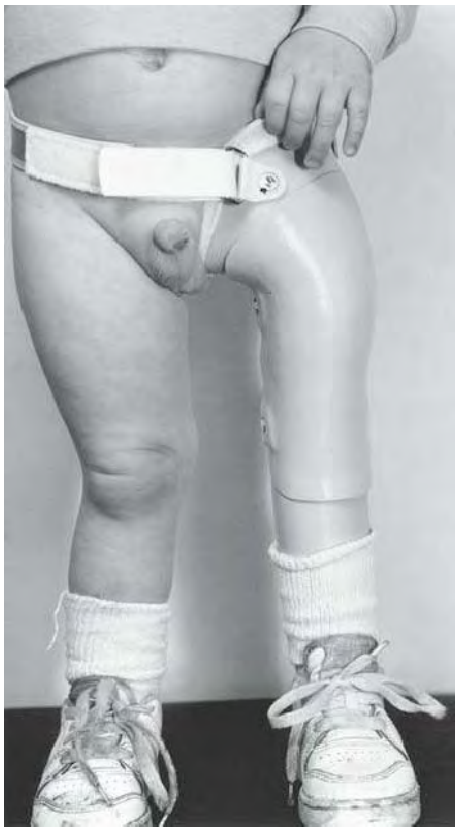
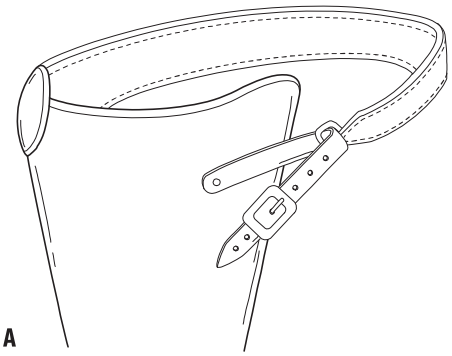


B

FIGURE 30-46. The nonconventional or extension prosthesis allows the child to "stand" on the prosthesis, extending his limb to the floor and accommodating the deformity. **A,B**: The nonconventional or extension prosthesis without a knee joint, which is usual. It is possible to add a knee joint to the prosthesis.

Suction sockets are not typically used for the pediatric population because of tight tolerances in fitting that cannot be maintained by a growing child. With the suction socket, the residual limb is pulled into a socket that incorporates a one-way valve in its design (Fig. 30-47). Once the valve is in place, the amputee expels air every time the prosthesis is in contact with the ground. During swing phase, the negative pressure within the socket holds the prosthesis in place. Air that leaks into the socket is quickly expelled through the one-way valve, and a constant negative pressure is maintained. Total contact suction sockets are generally used for the transfemoral amputee with a mature residual limb and at the completion of skeletal growth. Short limbs, volumetric changes, and severe scarring are contraindications for the suction-suspended socket.

Transtibial Prosthesis. The transtibial prosthesis is used the least in the pediatric population. Although most amputations in children are disarticulations, growth changes in the fibular deficiency often result in a transtibial-level residual limb that is a distal-end weight-bearing limb. The true transtibial socket is most often required for the traumatic amputee. Total contact design allows for increased pressure bearing over the patellar tendon, medial flare of the tibia, medial shaft of the tibia, and lateral shaft of the fibula, and the anterior and posterior compartments.



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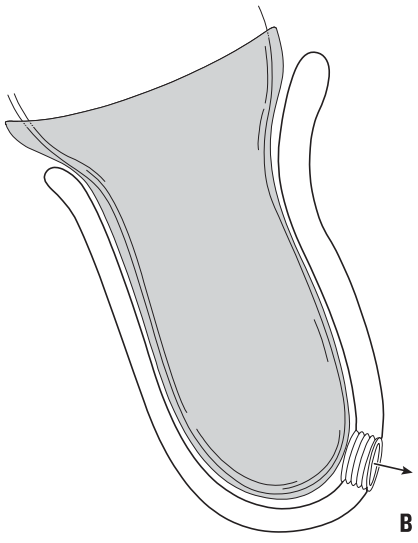


B

FIGURE 30-47. Methods of suspension for transfemoral prostheses. **A**: The Silesian belt is almost universally used in the young pediatric patient to suspend a transfemoral prosthesis or occasionally a knee disarticulation or transtibial prosthesis. **B**: The suction socket is a tight-fitting socket design with a one-way valve that allows air to be expelled with weight bearing to maintain a suction fit on the residual limb. It is best suited for the older child or adolescent, who has a mature limb that is not changing in size.

Similarly, the weight-sensitive areas most affected include the tibial crest, fibular head, distal tibia and fibula, peroneal nerve, and the patella. The socket design is composed of an outer shell, inner soft liner, and a cosmetic cover.

The patellar-tendon-bearing (PTB) socket is the standard socket (Fig. 30-48A). It provides the least suspension, and for that reason, is not often suitable for young children. The supracondylar/suprapatellar (SCSP) transtibial socket design (Fig. 30-48B) allows for suspension without the need for belts or cuffs. The medial, lateral, and anterior walls extend proximally, to fully enclose the patella and femoral condyles. The SC transtibial socket (Fig. 30-48C) is almost identical to

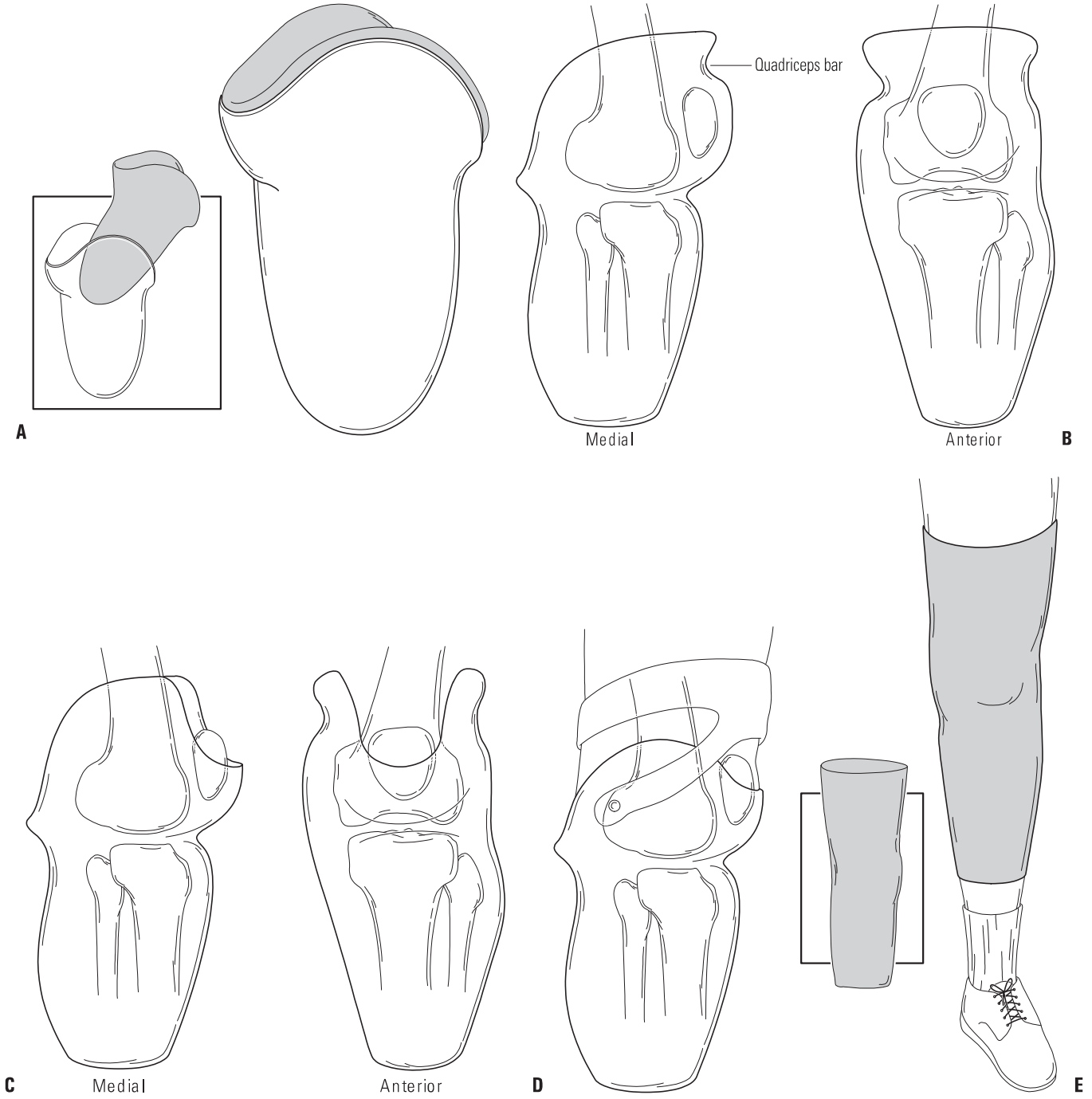


FIGURE 30-48. Common sockets and suspensions for transtibial pediatric amputees. **A**: PTB socket is most useful for the mature patient. It gives the most freedom of motion, but the least secure suspension. **B**: The PTB-SCSP design gives the most secure suspension and best knee control of any of the sockets that incorporate the suspension in the socket. **C**: The PTB-SC socket eliminates the suprapatellar portion of the socket anteriorly, providing better range of motion but less control of hyperextension. **D**: The SC cuff suspension is a common suspension used in the pediatric age range. **E**: The neoprene sleeve suspension provides very secure suspension for the very active amputee. This can also be used for the transfemoral amputee.

the SCSP design, except the anterior proximal brim does not enclose the patella, and therefore allows greater freedom and range of motion. Contraindications for both the SCSP and SC design include obese or muscular limbs and patients with heavy scarring around the knee.

SC cuff suspension is a common form of suspension for the pediatric transtibial amputee (Fig. 30-48D). The cuff is fabricated from leather and encompasses the femoral condyles and patella. It is attached to the medial and lateral aspects of the socket. The neoprene sleeve suspension is another useful suspension in the pediatric prosthesis (Fig. 30-48E). For the very active child, it provides a great level of security in that the prosthesis will not come off. Recent advances in silicone and urethane technology have increased comfort, flexibility, and cosmesis of the sleeve suspension systems.

Silicone suspension liners have become increasingly popular as a method of suspension without the need for belts or cuffs (Fig. 30-49). The liner is rolled onto the residual limb. At the distal end of the liner is a serrated pin. Inside the distal end of the socket is a shuttle or receptacle mechanism. Once the liner is donned, the amputee places the limb in the socket, and the pin and shuttle engage and lock into place. Pressing of a button hidden on the medial distal aspect of the prosthesis releases the pin, and the residual limb can be removed from the socket. Because of the physical characteristics of the liner, the greater the distracting forces placed on the prosthesis, the tighter the liner grips the residual limb. This system is used extensively in young children. Where space is at a premium, a cushioned silicone liner used in conjunction with a socket expulsion valve and a silicone sleeve allows the amputee to achieve a remarkable level of suspension using a modified suction technique.

Ankle Disarticulation Prosthesis (Syme). The obturator (medial opening) design is most often used when the distal bulbous end is large and the medial malleolus is prominent (Fig. 30-50A). The removable or segmented liner socket incorporates a full foam liner that has been built up to the same circumference as the distal bulbous end. A laminated shell is then formed over this insert. The patient dons the liner first, then slips this into the laminated receptacle (Fig. 30-50B). An atrophied residual limb with a small heel pad is best suited for this design, and the degree of cosmetic restoration will be very good. The silicone or bladder prosthesis utilizes an inner elastic area that stretches to permit the passage of the bulbous end of the residuum through the narrower circumference of the tibia and fibula, then constricts once the distal end has passed through

A

B

FIGURE 30-49. Silicone suspension liners (Triple S socket) have become very popular. The soft silicone liner has a serrated pin incorporated into the bottom of the liner. The patient rolls the liner on the residual limb **(A)**, then inserts the limb into the prosthesis **(B)**. At the bottom of the prosthesis is a socket in which the pin locks. It is released by pushing the button on the medial side of the prosthesis.

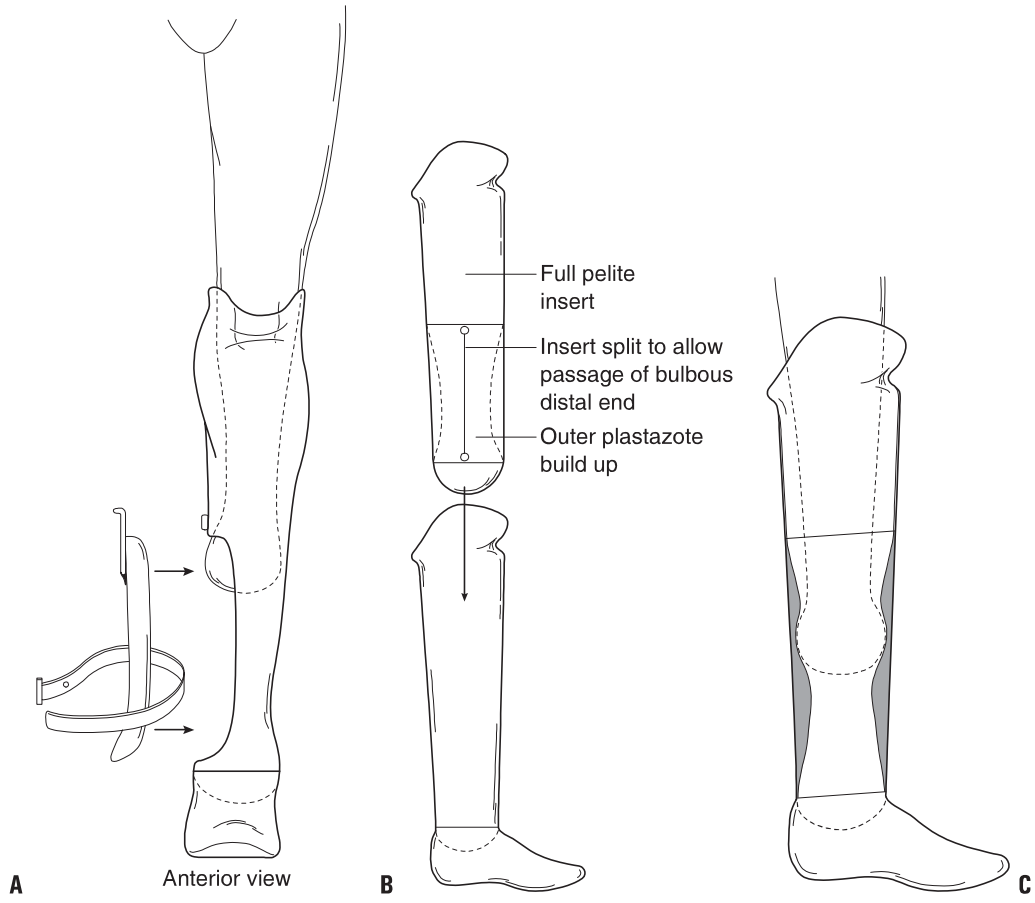


FIGURE 30-50. Common socket designs for Syme amputation prosthesis. **A**: Obturator design is often needed if the distal end of the residual limb is large and bulbous or the medial malleolus is prominent. It is the least cosmetic. **B**: The removable or segmented liner consists of a complete separate foam liner, which has a split in the side to allow the distal end of the limb to pass. Once the patient applies the liner, he or she slides the limb covered by the liner into the prosthesis. The patient must have the manual dexterity and strength to use this suspension, which will eliminate some patients with hand anomalies from using it. Limbs with a small heel do best with this system. **C**: The bladder design has a built-up silicone sleeve, which the patient slides the limb past. This socket fits more loosely and does not stabilize the heel pad as well as the other designs.

(Fig. 30.50C). All of the above designs maintain total contact, and the proximal brim is at the level of the patellar tendon. This ensures that the biomechanical forces are adequately spread up to a load-bearing landmark to increase comfort and function.

Prosthetic Knees. It has been estimated that more than 100 prosthetic knees are commercially available, and the number is growing each year (223). Although most are for adults, recently there are a number of new knees available for children. The prosthetic knee is composed of the knee mechanism or frame and may contain a control unit. The control unit consists of a pneumatic, hydraulic, or mechanical system, or some combination of these three. The control unit responds to changes in cadence and dampens sudden, abrupt changes. The faster a hydraulic or pneumatic unit is compressed, the faster the energy is released, and this helps to regulate the lower shank of the prosthesis. The prosthetic knee unit can be further subdivided into single axis and polycentric types.

The maturation of gait from infant to adult carries with it the need for sound practice in selecting the appropriate knee, on the basis of amputation level, functional level, and body size. In general, the single-axis internal knee without any control unit is the first knee to be used on the child, because of its light weight, short lever arm, and simplicity. In the single-axis knee joint, the lower shank rotates around a single point in relation to the socket.

A polycentric knee was introduced in 1998 for the infant and toddler, and it may be used if space permits. Internal polycentric knees move around a center of rotation that varies with the flexion angle of the lower shank (213). The four-bar linkage knee is the most common polycentric knee and the most widely used by prosthetists (224) (Fig. 30-51). The inherent stability during stance, the fluid knee-flexion movement, and the mechanical design to give more ground clearance during flexion increase patient and practitioner confidence in the unit (225).

Changes in design and technology have now widened the boundaries and age distinctions for the prescribing of specific

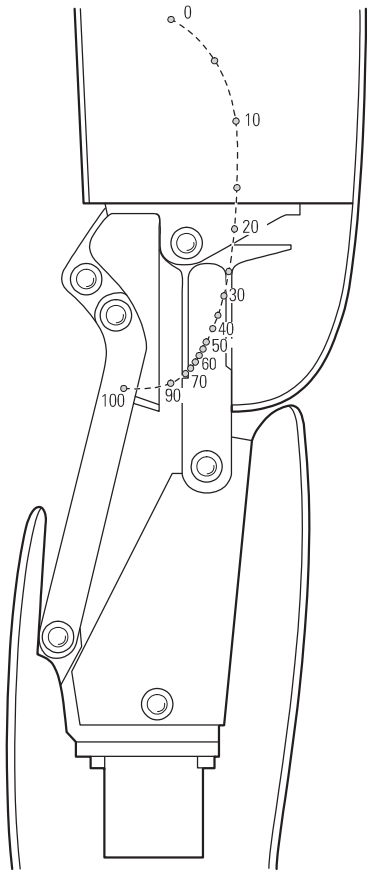


FIGURE 30-51. Four-bar linkage is an internal polycentric knee that provides many advantages to the patient, including increased stability and better ground clearance during swing phase. As indicated in this illustration, the point of rotation varies with the degree of flexion. With the knee flexed, the leg folds under the thigh segment and therefore is very useful for longer residual limbs. There is also a hydraulic version of the four-bar linkage for children.

knees. Traditionally, an articulating knee would be introduced in a congenital amputee at approximately 3 to 4 years of age. This age was determined, in part, by the limitations in the size and function of the components. In the experience at the authors' center, as well as others, introduction of a prosthetic knee without a locking feature can be used as the first prosthesis when the child first pulls to stand. A recent report demonstrated the benefits of early fitting with articulated knees in children as young as 17 months. All children learned to walk with an articulated knee, despite their age differences (157).

As the child develops and grows, more sophisticated control systems (e.g., hydraulic knees) can be incorporated into the prosthesis. Most components carry specific weight guidelines, and many children reach these ranges well before adulthood. For example, an adult hydraulic polycentric knee is routinely used on 8-year-old boys whose weight has surpassed 100 lb. This does not mean that every child of a certain age and weight should have a particular knee. Placing a sophisticated knee and control system on an individual who has neither the hip range, muscle strength, nor residual limb length to activate the knee often results in contralateral hip and lower back pain as well as patient frustration.

Prosthetic Feet. Variations in the materials, design, and alignment of the foot can have profound effects on the performance of the prosthesis. Functionally, prosthetic feet can be categorized into five main groups (226):

- Solid ankle cushion heel (SACH)
- Single axis
- Multiaxis
- Elastic keel
- Dynamic response

The SACH foot [Therapeutic Recreation Systems, Inc. (TRS)] contains no articulating parts, and foot motion depends on the various compressive properties of the materials used between heel-strike and toe-off (Fig. 30-52A). It is generally considered when amputees require maximum late-stance stability because of weak knee extensors, knee-flexion contractures, or poor mid- to late-stance balance (227). The SACH foot is used in pediatrics when the foot size is below 12 cm. The Little Feet is a bolt on type SACH foot designed with unique energy dynamics (Fig. 30-52B). The toes are very flexible because of the use of an elastomer that more closely mimics the child's foot. A special removable heel core allows the foot to be used "barefoot."

The single-axis foot usually contains rubber bumpers that allow passive dorsi- and plantar flexion. By changing the hardness of the bumper, the prosthetist is able to effectively change the properties of the foot. The single-axis foot does not come in a size suitable for the child amputee. This foot is best suited for the transfemoral amputee, in whom full-foot contact with the ground is necessary to increase stability. The multiaxis foot allows passive dorsi- and plantar flexion, inversion, and eversion. The multiaxis foot was once thought best suited for the amputee who because of uneven terrain or a lifestyle that includes golfing or various sports requires flexibility and some rotational control: It has now found its way into the pediatric population. The multiaxial foot (College Park Truper foot) (Fig. 30-52C) allows controlled resistance in all planes—inversion/eversion, dorsiflexion and plantar flexion, and transverse rotation about the ankle joint. This class of foot has gained wide acceptance within the pediatric arena, in part because of its ability to absorb forces at the ankle and reduce transmission of these forces to the socket. This is particularly useful when fitting a very short residual limb.

Once the child's activity level warrants a higher functioning foot, the prosthetist can move the child into a dynamic-response foot. This group of feet is distinguished by a spring mechanism in the keel that deflects during gait (Fig. 30-52D). The dynamic-response foot has found its way into competitive-level sports as well as day-to-day activities. Although the variety of componentry for children is still much less than for adults, there is a wide variety of feet with different performance characteristics available. It is important to use components that will maximize performance and at the same time be appropriate for the patient (228).

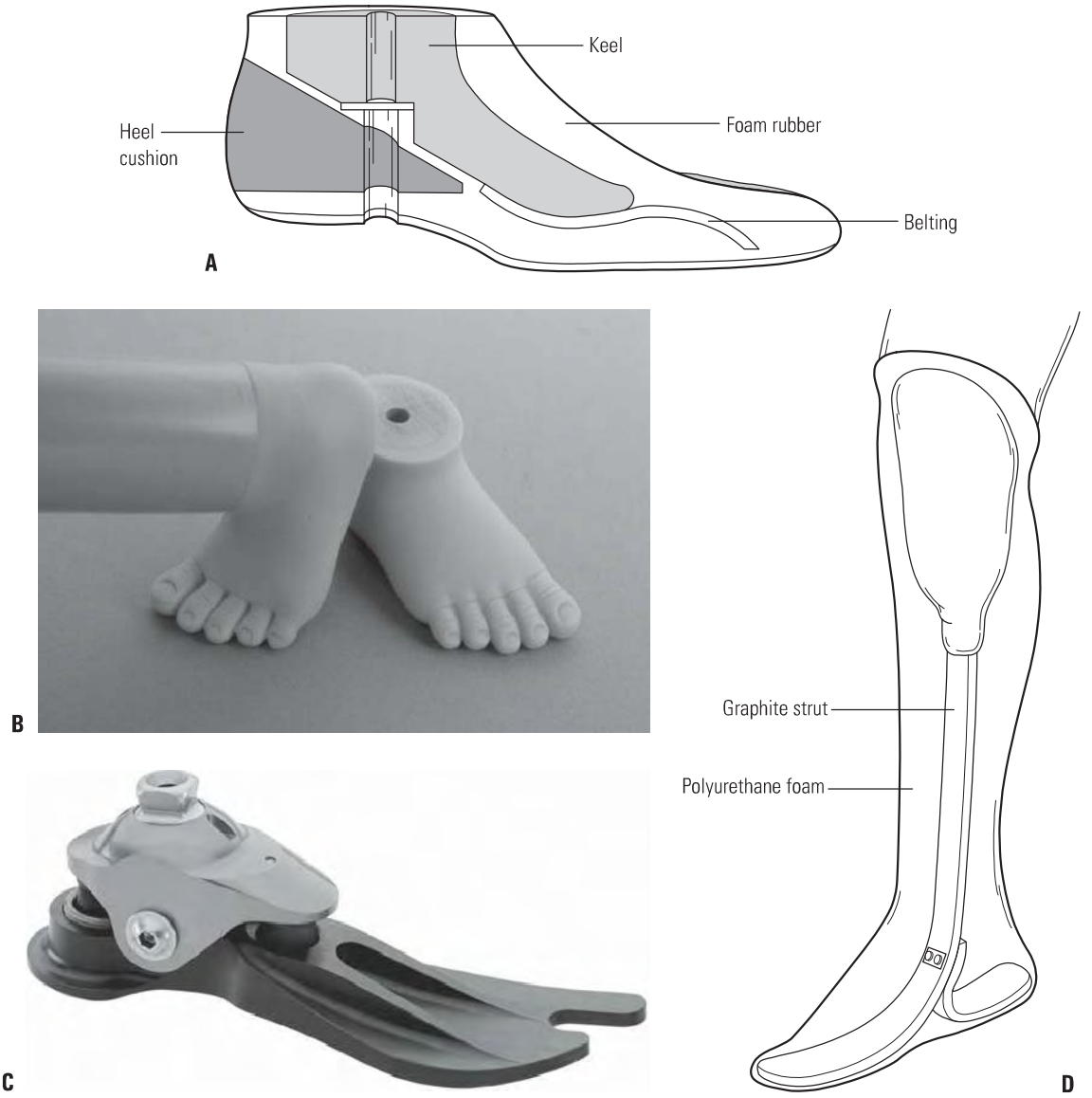


FIGURE 30-52. **A**: In the conventional style SACH foot, the length of the keel controls the toe lever arm, and therefore the hyperextension moment at the knee, while the compression of the material at the heel absorbs the forces at heel strike. **B**: The Little Feet type design incorporates unique energy dynamics with flexible toes all in sizes beginning at 10-cm length. The length of the keel controls the toe lever arm and thus the hyperextension moment at the knee, and the compression of the elastomer heel absorbs and deflects the forces at heel strike. **C**: The dynamic multiaxial TruPer foot allows rotation, inversion and eversion, flexion, and extension movements. The outer cosmetic shell can be exchanged for larger shells as the child grows. **D**: The Flex-foot is a dynamic-response foot with much different performance characteristics than the SACH foot or multiaxial foot. It is used for the older, stronger, and physically active child who has the physical ability to use such a foot.

At slower walking velocities, there is little difference between the dynamic-response foot and the SACH foot (229). Generally, children are not fitted with the highest-performance dynamic-response feet, because of constant growth and weight changes and the high costs associated with foot replacement. The involvement in competitive sports is usually a good benchmark to initiate fitting adolescents with the highest-performance dynamic-response feet.

In the transfemoral amputee, it is crucial to properly choose a foot that will enhance gait, but also to choose the complementary knee that will aid in controlling the foot during all aspects of the gait cycle. A common mistake is to prescribe a dynamic-response foot with a simple, friction-controlled knee that is incapable of preventing uncontrolled heel rise. The same is true of the transtibial amputee, who lacks the muscle strength to control the foot, often resulting in premature muscle fatigue. In the selection of multiple components, the prosthetist must marry the characteristics of all components, so that maximum benefit can be available to the amputee.

Partial-foot Prosthesis. The most important consideration in the fitting of the partial-foot amputee is to ensure that adequate load-bearing is designed into the prosthesis of choice. As a general rule, the more proximal the level of amputation, the higher the prosthesis must fit over the ankle complex and the more proximally it must fit on the tibia and fibula. Tissue condition, function of the remaining foot complex, and activity of the child all play a role in determining the prescription and design of the prosthesis.

Complete or partial absence of the toes usually requires little more than a shoe filler. A carbon fiber insert to better control forces from heel to toe-off may be incorporated in the shoe filler. In the case of the very young child, no intervention may be required until a need has been demonstrated, for example, the inability to keep the shoe on, especially when the child becomes more active in sports.

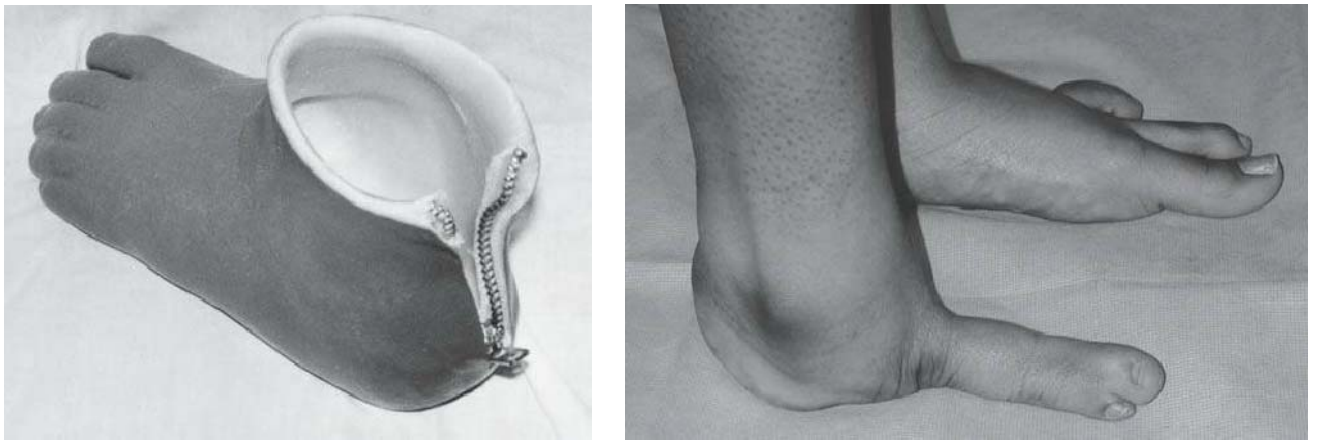
The prosthesis most commonly used for the moderate/short partial-foot amputee is the Lange silicone partial-foot prosthesis (Fig. 30-53). This incorporates a cosmetic foot shell, silicone-laminated socket with modified foot sole, and a posterior zipper for ease of donning and doffing. The prosthesis is fabricated over a modified model of the patient's partial foot. The socket trim line is proximal to the malleoli and is fitted intimately to ensure adequate control. The design of a partial-foot prosthesis may also include a removable insert, to accommodate the need for corrective alignment of the residual foot. The prosthesis is then cosmetically finished to resemble the contralateral limb. Overall, this type of prosthesis is perfectly suited for the child amputee and resists premature wear and tear. If needed, a partial-foot prosthesis should be prescribed once the child is pulling to furniture, so that foot control will begin at an early age. It should be noted that a low-profile insert (distal to the malleoli), used in conjunction with a high-top boot, will offer adequate function and cosmesis until a lower cut shoe is requested by the parent.

The Chopart, or midtarsal, amputation is rarely used except in special instances (230). In the Chopart partial-foot amputation level, the prosthesis is modified to encompass the calcaneus and talus, and this results in a prosthesis that is often longer than the contralateral limb. The prosthesis must encompass the ankle joint, and it often rises proximally to the patellar tendon in an effort to reduce forces on the tibial crest–socket interface. Selection of prosthetic feet is compromised because of the lack of space distally, and commercially available carbon foot plates require permanent attachment with vulcanizing rubber cement. This negates any changes caused by growth, and realignment to compensate for gait changes is virtually impossible.

Terminal Devices for the Upper Extremity. The choices left open to the prosthetist are numerous and, at times, controversial. Where some clinics maintain rigid protocols for terminal device selection, other clinics rely more on patient and parent input, combined with historic success rates for device types. Clinics that maintain very high caseloads for myoelectric devices, for example, will most likely have far more experience in fitting externally powered prostheses, compared to a clinic that may only see a handful of potential myoelectric candidates.

In simple terms, the terminal devices can be divided into hands and hooks, and they can be body powered (cable and harness) or externally powered (electric). Hands and hooks can be either voluntary opening or voluntary closing. Patton lists the functional and prescription criteria for the various terminal devices (203).

The initial fitting of a child with upper extremity limb deficiency begins at 4 months of age in a passive prosthesis with a stylized passive hand. There are several hands manufactured for this age range (Fig. 30-54A). This allows for equal arm lengths for the development of propping up on the amputated side and greater acceptance by the parents. Following initial sitting balance, the clenched-fist terminal device is exchanged for a small infant passive hand. When the infant begins to reach out (at ~15 to 18 months of age), the clinic team begins to assess the need for either body-powered or externally powered prostheses. If body-powered prosthesis is recommended, a cable-operated Hosmer 12P plastic-covered hook (Fig. 30-54B) or an ADEPT infant hand (Fig. 30-54C) will be



A B

FIGURE 30-53. **A**: The Lange silicone partial-foot prosthesis is a custom-made prosthesis that can incorporate a keel to aid in foot stability and push-off in gait. **B**: It is useful for children with partial amputations of the foot or congenital longitudinal deficiencies of the foot, shown here. It is not useful in feet with insufficient length, for example, those with the Chopart amputation.

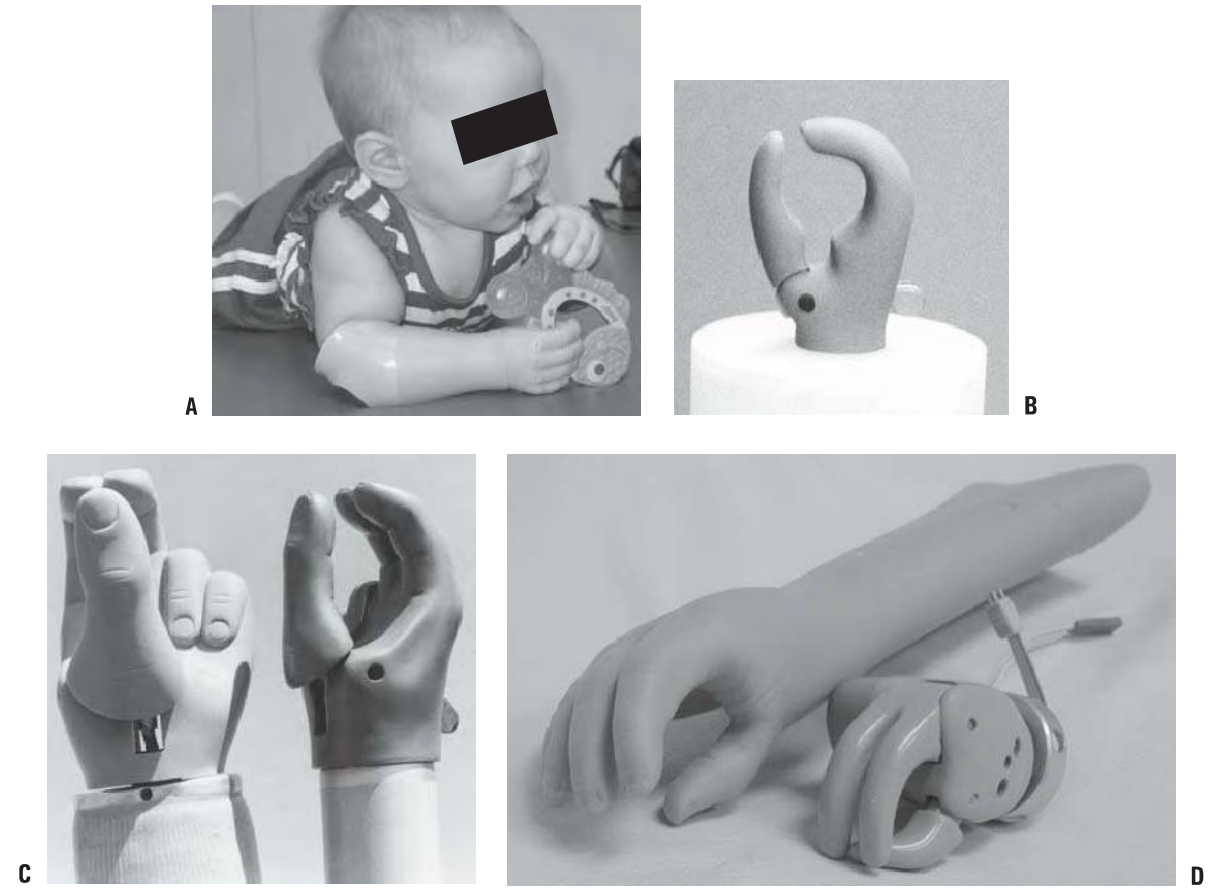


FIGURE 30-54. **A**: The TRS ALPHA Infant Hand is an option for the first prosthetic hand offering age-appropriate fine motor activities. This would commonly be fitted between 4 and 6 months of age. The hand incorporates a flexible thumb that allows objects to be placed for simple grasp and release functions. **B**: The ADEPT is a voluntary closing body-powered hook that is fitted at approximately 15 months of age, if the child is ready and a body-powered device is desired. **C**: The Lite-Touch is a voluntary closing hand that would find the same indications as the ADEPT hook. It looks a bit more like a hand, which often makes this option popular with parents. **D**: The Variety Village hand is one of the most commonly used myoelectric hands in the pediatric age group. The Otto Bock Electrohand is shown in Figure 30-42. Both of these electric hands are covered with a cosmetic glove.

prescribed. The canted design of the 12P hook allows for greater visual feedback to the wearer. In the event that a myoelectric device is warranted, the Variety Village 0-3 (VV 0-3) electric hand (Fig. 30-54D) or the Otto Bock Electrohand (Fig. 30-42) is used. In the pediatric VV 0-3 hand, the thumb and opposing two fingers operate to form a three-point chuck grip. In the Otto Bock 2000 hand, the same principle is applied, except that from the open to closed position, the thumb sweeps from a lateral position to meet the two opposing fingers upon close.

Progression from this starting point through the various component sizes and versions allows for a relatively smooth transition into adulthood. Prescription criteria are reviewed during each clinic visit, and changes are made on the basis of the child's changing needs. In today's environment of active children and sports activities, the use of sports or other adaptive terminal devices is essential for the amputee. TRS has developed numerous devices for use in sports and recreational activities. These can be interchanged on the prosthesis, so that only one socket is required.

Endoskeletal versus Exoskeletal Construction. The structure or construction of a prosthesis is referred to as an endoskeletal (internal structure) or exoskeletal (external structure) prosthesis. Generally, transtibial, partial foot, and transradial prostheses are constructed exoskeletally, and transfemoral, knee disarticulation, hip disarticulation, transhumeral, and shoulder disarticulation levels of prostheses are constructed endoskeletally.

Exoskeletally finished prostheses are more durable and better suited to the growing child. There are various techniques and materials used in the construction of the exoskeletal prosthesis. Generally, following the completion of dynamic alignment, the ready-to-be-finished prosthesis is placed within a transfer jig that allows the socket to be separated from the foot while maintaining alignment. A rigid polyurethane foam is added, and the prosthesis is cosmetically shaped to equal the sound limb. The structure is then laminated with acrylic resin forming the outer "shell." The advantages of this construction are its increased durability and that it is easy to clean and structurally strong. The major disadvantages are the lack

of further alignment capabilities and that they are less cosmetically acceptable than some other types.

Endoskeletal design was initially used in the immediate postoperative period as a temporary method to initiate ambulation while maintaining the ability to alter the alignment. This quickly became the norm for fitting in the adult population and has been used primarily for knee disarticulation or transfemoral prosthesis. The prosthesis is modular and composed of a pylon (tube) and connecting hardware, and it allows for quick changing of damaged components. In the event that realignment is necessary, the endoskeletal design incorporates alignment jigs within the attachment couplings, and only the cosmetic soft cover needs to be removed for adjustment. For these reasons, advanced components for use by children tend to be engineered for use in this system. The disadvantages of the endoskeletal system are lack of durability of the cosmetic cover, increased maintenance, and increased costs. This is outweighed by the increase in function and ease of adjusting length.

ROLE OF THE PHYSICAL/OCCUPATIONAL THERAPIST

The role of the physical/occupational therapist in the care of the limb-deficient child is mostly nontraditional. In addition to the traditional role, the physical/occupational therapist fills the roles of teacher, advocate, friend, and liaison (231). In some situations, all but the traditional role of therapist may be filled by a nurse. In some cases, the role may be shared. What is important is to recognize the need for all of these activities.

The first role of the therapist will usually be that of the educator. In this role, it is important that the therapist, physician, and prosthetist all be of one mind regarding the patient's treatment. First is the education of the parents. Like nurse-practitioners, the therapists will usually have more time and be better heard than the doctor when relaying information to the parents about their child. The therapist will be able to reinforce to the parents the options that have been discussed at the initial meeting with the physician. Most importantly, they can arrange for the family and child to meet others with similar deficiencies during routine sessions. Throughout the child's life, the experienced therapist can be of immense value to the child and young adult in anticipating problems and helping with solutions.

The therapist's first role as advocate will start with the first meeting with the parents. The importance in this role is to bring the parents to see the normal, as well as the abnormal, and to ensure that the initial bonding to the parents occurs. The therapist will frequently need to advocate for the patient to insurance companies and other agencies to help provide for the patient's needs. When the child starts into day care and then school, the therapist will assist in the child's transition into a new world by educating the teachers and the child's peers about the child's differences. This can be extremely important for the child's acceptance and socialization. Later, the same role may be necessary with physical education teachers and coaches to ensure that the child can participate in all the activities he or she is able to.

The therapist (or nurse) with these roles is the ideal liaison among the team members. This close teamwork can spare the child and parents countless visits to clinics and delays in treatment: a very important goal in avoiding the medicalization of a condition for which there is no cure, but only good management.

The traditional role of the therapist will be far more home/community based than hospital/office based for many reasons. First, the child's condition will be permanent. This means adapting to the environment in which the child exists. None of the child's activities (toileting, eating, dressing, play, sports) will take place in the hospital or an office. Therefore, they should be learned in the normal environment. Second, the parents will be with the child and are responsible for the child's development and learning. They will have unlimited access to the child for this "therapy." Finally, the child should not come to think of himself or herself as a medical problem, but rather as a child with a difference that can be successfully adapted to. Unnecessary hospital, clinic, or office visits are not a good way in which to communicate this goal of independence.

The traditional medical model does have a role in acute situations, as it does in many diseases or postsurgical situations. These are times when specific therapeutic exercises or the use of new prostheses must be performed, supervised, and taught. During the first months of life of a congenital amputee, the parents are seen by the therapist every 6 to 8 weeks. The child's development is monitored, and the parents are taught activities appropriate for the stage of development, for example, rolling over and coming to stand from sitting. These visits are used to monitor the parents' coping, to listen and answer questions, review treatment options, and arrange for the meeting with other parents and children.

Following surgical intervention and prosthetic fitting, the medical model is more appropriate, and the frequency of the visits increases briefly, while the child and parent are taught the use of the prosthesis. In addition to the usual goals of increasing or maintaining motion and strength following surgery, the therapist plays a critical role in edema control. This is very important if the patient is to resume prosthetic wear quickly. Teaching and supervising elastic wrapping and obtaining shrinkers are important postsurgical issues.

Older children need to be taught ways to become independent in donning and doffing the prosthesis, toileting, and other activities with the prosthesis. Fitting of a new prosthetic component, for example, a hydraulic knee joint, will usually require specific training to maximize the benefits of the new components.

As the child grows out of infancy, the therapist can be helpful in designing and modifying age-appropriate play activities, With the approach of school age, emphasis switches to independence in the ADL, and later, to fine motor skills that may be needed for classroom activities. Adaptations for sports, for example, special terminal devices, if desired, or a swimming leg, are important, as is the advocacy role to allow the children to participate in all possible activities.

In adolescence, the need for specific therapeutic interventions is usually minimal. The child has now become fully aware of his or her differences and their significance. The child

will usually dictate the needs. Appearance being important, more cosmetic prostheses and improved gait become important issues.

At this time, driving becomes one of the major issues defining independence. The therapist can play a critical role in directing the child and parents to the appropriate agency for the rules of the state, and to a source for evaluation and modifications to the vehicle. As with any adolescent, the amputee should attend driver education training, using modifications if needed. Modifications can range from simple to complex. Switching the brake and gas pedals to accommodate unilateral lower limb loss is one of the most common examples. Many amputees, even with bilateral lower extremity loss, drive without adaptations. This must be closely monitored and evaluated by the state's examiners. Hand controls are used most commonly in bilateral lower extremity loss. A ring adaptation can be used to modify the steering wheel for upper extremity amputees. A handicap license is appropriate for individuals whose mobility is limited. The higher level amputee, and those with multilevel limb loss, may benefit from a handicap parking license. All modifications are listed on the amputee's driver's license.

College is often the patient's first test of complete independence, and the needs may be greater than what the child anticipates. The therapist can be of great value in assessing the situation, counseling the patient, and helping with the transition. The Internet can prove to be a great resource in assisting college-bound students and their families.

ADAPTATIONS FOR ACTIVITIES AND SPORTS

Children of all ages with limb deficiencies or amputations should be encouraged to participate in sports and recreational activities with their peers. The psychological impact of sports cannot be underestimated. Improving self-esteem and confidence, gaining independence, learning to win and lose, developing decision-making and problem-solving skills, and cooperating as a team member are a few of the benefits that a child carries throughout his or her life. Improving physical fitness, developing balance, strength, coordination and motor skills, increasing endurance, and weight control are benefits of physical activity.

Over the years, there has been an increased awareness of adapted sports and recreation for individuals with physical and mental impairments. The Paralympic and Special Olympics initiatives have been the most obvious and have sparked an increase in availability of programs for special-needs children. Laws also have been passed for children to receive education in the least restrictive environments. PL 94-142 provides free and appropriate education for all children with disabilities (231). Physical therapy and recreation are related services included in this legislation. This allows for adapted physical education to be included in a child's everyday school activities. Physical therapists need to be aware of the resources and adaptations that provide accessibility of a sport to a child within limits of his/her physical abilities in the school and community settings. Gross motor achievements are a measure of a child's development. Learning to toss a ball, jump, run, hop, and ride a bicycle are activities included on standardized developmental screenings and tests. It is important that these activities be included in the child's plan of treatment, so that a child can be offered the same age-appropriate physical challenges as his or her peers.

Special adaptations and specific sports-related prostheses are available, depending on the degree and level of the disability (Fig. 30-55). These adaptations are too numerous to mention. Recreational and sports-related terminal devices are available for the upper extremity amputee (232, 233). Adaptations can be as simple as raising the handlebars on a bicycle and adding a toe strap to highly sophisticated prosthetic components specific to each sport (234). Information and resources for sports and adaptive recreation for the amputee can be obtained through the Amputee Coalition of America (www.amputee-coalition.org; telephone 1-888-267-5669). The Association of Children's Prosthetic and Orthotics Clinics is an excellent resource for services in geographical regions (www.acpoc.org; telephone 847-698-1637).

FIGURE 30-55. As seen in this photograph, participation in high-level sports is possible for children and adolescents with limb deficiency. Advances in prosthetic modifications design and national organizations that promote athletics give patients the freedom to pursue a wide array of winter and summer sports.

REFERENCES

1. Frantz CH, O'Rahilly R. Congenital skeletal limb deficiencies. *J Bone Joint Surg Am* 1961;43:1202–1224.
2. Schuch C, Pritham CH. International forum: International Standards Organisation terminology: application to prosthetics and orthotics. *J Prosthet Orthot* 1994;6:29–33.
3. Krebs DE, Fishman S. Characteristics of the child amputee population. *J Pediatr Orthop* 1984;4:89–95.
4. Akinyoola AL, Oginni LM, Adegbehingbe OO, et al. Causes of limb amputations in Nigerian children. *West Afr J Med* 2006;25:273–275.
5. Froster-Iskenius UG, Baird PA. Limb reduction defects in over one million consecutive livebirths. *Teratology* 1989;39:127–135.
6. Stewart CP, Jain AS. Congenital limb anomalies and amputees Tayside, Scotland 1965–1994. *Prosthet Orthot Int* 1995;19: 148–154.
7. Brown N, Lumley J, Tickle C, et al. *Congenital limb reduction defects*. London, UK: Department of Health, The Stationery Office, 1996.
8. Evans JA, Vitez M, Czeizel A. Congenital abnormalities associated with limb deficiency defects: a population study based on cases from the Hungarian Congenital Malformation Registry (1975–1984). *Am J Med Genet* 1994;49:52–66.
9. Fixsen JA. Major lower limb congenital shortening: a mini review. *J Pediatr Orthop B* 2003;12:1–12.
10. Bavinck JN, Weaver DD. Subclavian artery supply disruption sequence: hypothesis of a vascular etiology for Poland, Klippel-Feil, and Mobius anomalies. *Am J Med Genet* 1986;23:903–918.
11. Streeter GL. Focal deficiencies in fetal tissues and their relation to intrauterine amputation. *Contrib Embryol* 1930;22:1–44.
12. Glessner JR. Spontaneous intrauterine amputation. *J Bone Joint Surg Am* 1963;45:351–355.
13. Bamforth JS. Amniotic band sequence: Streeter's hypothesis reexamined. *Am J Med Genet* 1992;44:280–287.
14. Sadler TW. Embryology and gene regulation of limb development. In: Herring JA, Birch JG, eds. *The child with a limb deficiency*. Rosemont, IL: American Academy of Orthopaedic Surgeons, 1998:3–12.
15. Lovett MG, Clines G, Wise CA. Genetic control of limb development. In: Herring JA, Birch JG, eds. *The child with a limb deficiency*. Rosemont, IL: American Academy of Orthopaedic Surgeons, 1998:13–24.
16. Lee GS, Kochhar DM, Collins MD. Retinoid-induced limb malformations. *Curr Pharm Des* 2004;10:2657–2699.
17. Skjaerven R, Wilcox AJ, Lie RT. A population-based study of survival and childbearing among female subjects with birth defects and the risk of recurrence in their children. *N Engl J Med* 1999;340:1057–1062.
18. Weaver DD. Vascular etiology of limb defects: the subclavian artery supply disruption sequence. In: Herring JA, Birch JG, eds. *The child with a limb deficiency*. Rosemont, IL: American Academy of Orthopaedic Surgeons, 1998:25–38.
19. Holmes LB. Teratogen-induced limb defects. *Am J Med Genet* 2002;112:297–303.
20. Kübler-Ross E. *On death and dying*. New York, NY: Macmillan, 1969.
21. Letts M, Vincent N. Congenital longitudinal deficiency of the fibula (fibular hemimelia). Parental refusal of amputation. *Clin Orthop* 1993;287:160–166.
22. Kerr SM, McIntosh JB. Coping when a child has a disability: exploring the impact of parent-to parent support. *Child Care Health Dev* 2000;26: 309–322.
23. Dunn NL, McCartan KW, Fuqua RW. Young children with orthopedic handicaps: self-knowledge about their disability. *Except Child* 1998;55:249–252.
24. Vaughn BE, Langlois JH. Physical attractiveness as a correlate of peer status and social competence in preschool children. *Dev Psychol* 1983;19:561–567.
25. Harper DC, Wacker DP, Cobb LS. Children's social preferences toward peers with visible physical differences. *J Pediatr Psychol* 1986;11:323–342.
26. Richardson S. Age and sex differences in values toward physical handicaps. *J Health Soc Behav* 1970;11:207–214.
27. Richardson S. Handicap, appearance and stigma. *Soc Sci Med* 1971;5: 621–628.
28. Varni JW, Setoguchi Y, Rappaport LR, et al. Effects of stress, social support, and self-esteem on depression in children with limb deficiencies. *Arch Phys Med Rehabil* 1991;72:1053–1058.
29. Varni JW, Setoguchi Y. Correlates of perceived physical appearance in children with congenital/acquired limb deficiencies. *J Dev Behav Pediatr* 1991;12:171–176.
30. Varni JW, Setoguchi Y. Effects of parental adjustment on the adaptation of children with congenital or acquired limb deficiencies. *J Dev Behav Pediatr* 1993;14:13–20.
31. Hubbard S, Heim W, Giavedoni B. Paediatric prosthetic management. *Curr Orthop* 1997;11:114–121.
32. Amstutz HC. The morphology, natural history, and treatment of proximal femoral focal deficiency. *A symposium on proximal femoral focal deficiency—a congenital anomaly*. Washington, DC: National Academy of Sciences, 1969:50–76.
33. Amstutz HC. Natural history and treatment of congenital absence of the fibula. *J Bone Joint Surg Am* 1972;54:1349.
34. Hamanishi C. Congenital short femur: clinical, genetic and epidemiological comparison of the naturally occurring condition with that caused by thalidomide. *J Bone Joint Surg Br* 1980;62:307–320.
35. Achterman C, Kalamchi A. Congenital deficiency of the fibula. *J Bone Joint Surg Br* 1979;61:133–137.
36. Westin GW, Sakai DN, Wood WL. Congenital longitudinal deficiency of the fibula: follow-up of treatment by Syme amputation. *J Bone Joint Surg Am* 1976;58:492–496.
37. Anderson M, Messner MB, Green WT. Distribution of lengths of the normal femur and tibia in children from one to eighteen years of age. *J Bone Joint Surg Am* 1964;46:1197–1202.
38. Christie J, Lamb DW, McDonald JM. A study of stump growth in children with below-knee amputations. *J Bone Joint Surg Br* 1979;61: 464–465.
39. Aitken GT. Overgrowth of the amputation stump. *Interclin Inform Bull* 1962;1:1–8.
40. Jorring K. Amputation in children: a follow-up of 74 children whose lower extremities were amputated. *Acta Orthop Scand* 1971;42:178–186.
41. Pellicore RJ, Sciora J, Lambert CN, et al. Incidence of bone overgrowth in the juvenile amputee population. *Interclin Inform Bull* 1974;8:1–8.
42. O'Neal ML, Bahner R, Ganey TM, et al. Osseous overgrowth after amputation in adolescents and children. *J Pediatr Orthop* 1996;16:78–84.
43. Aitken GT. Surgical amputation in children. *J Bone Joint Surg Am* 1963;45:1735–1741.
44. Speer DP. The pathogenesis of amputation stump overgrowth. *Clin Orthop* 1981;159:294–307.
45. Abraham E. Operative treatment of bone overgrowth in children who have an acquired or congenital amputation. *J Bone Joint Surg Am* 1996;78:1287–1288.
46. Davids JR, Meyer LC, Blackhurst DW. Operative treatment of bone overgrowth in children who have an acquired or congenital amputation. *J Bone Joint Surg Am* 1995;77:1490–1497.
47. Tenholder M, Davids JR, Gruber HE, et al. Surgical management of juvenile amputation overgrowth with a synthetic cap. *J Pediatr Orthop B* 2004;24:218–226.
48. Benevina J, Makley JT, Leeson MC, et al. Primary epiphyseal transplants and bone overgrowth in childhood amputations. *J Pediatr Orthop* 1992;12:467–450.
49. Bernd L, Blasius K, Lukoschek M, et al. The autologous stump plasty: treatment for bony overgrowth in the juvenile amputees. *J Bone Joint Surg Br* 1991;73:203.
50. Bickels J, Kollender Y, Pritsch T, et al. Knee stability after resection of the proximal fibula. *Clin Orthop Relat Res* 2007;454:198–201.
51. Einoder PA, Choong PF. Tumors of the head of the fibula: good function after resection without ligament reconstruction in 6 patients. *Acta Orthop Scand* 2002;73(6):663–666.
52. Malawer MM. Surgical management of aggressive and malignant tumors of the proximal fibula. *Clin Orthop Relat Res* 1984;(186):172–181.

53. Innocenti M, Delcroix L, Manfrini M, et al. Vascularized proximal fibular epiphyseal transfer for distal radial reconstruction. *J Bone Joint Surg Am* 2004;86(7):1504–1511.
54. Innocenti M, Ceruso M, Manfrini M, et al. Free vascularized growth-plate transfer after bone tumor resection in children. *J Reconstr Microsurg* 1998;14(2):137–143.
55. Bowen RE, Struble SG, Setoguchi Y, et al. Outcomes of lengthening short lower-extremity amputation stumps with planar fixators. *J Pediatr Orthop* 2005;25(4):543–547.
56. Bernstein RM, Watts HG, Setoguchi Y. The lengthening of short upper extremity amputation stumps. *J Pediatr Orthop* 2008;28(1):86–90.
57. Alekberov C, Karatosun V, Baran O, et al. Lengthening of congenital below-elbow amputation stumps by the Ilizarov technique. *J Bone Joint Surg Br* 2000;82:239.
58. Tooms R. Acquired amputations in children. In: Bowker J, Michael EJ, eds. *Atlas of limb prosthetics: surgical, prosthetic, and rehabilitation principles*. St. Louis, MO: Mosby, 1992:738.
59. Wilkins K, McGrath P, Finley G, et al. Phantom limb sensations and phantom limb pain in child and adolescent amputees. *Pain* 1998;78:7.
60. Melzack R, Israel R, Lacroix R, et al. Phantom limbs in people with congenital limb deficiency or amputation in early childhood. *Brain* 1997;120:1603.
61. Lindesay J. Multiple pain complaints in amputees. *J R Soc Med* 1985;78:452.
62. Davis RW. Phantom sensation, phantom pain and stump pain. *Arch Phys Med Rehabil* 1993;74:79.
63. Krane EJ, Heller LB. The prevalence of phantom sensation and pain in pediatric amputees. *J Pain Symptom Manage* 1995;10(1):21–29.
64. Ong BY, Arneja A, Ong EW. Effects of anesthesia on pain after lower-limb amputation. *J Clin Anesth* 2006;18(8):600–604.
65. Wilson JA, Nimmo AF, Fleetwood-Walker SM, et al. A randomised double blind trial of the effect of pre-emptive epidural ketamine on persistent pain after lower limb amputation. *Pain* 2008;135(1–2):108–118.
66. Grant AJ, Wood C. The effect of intra-neural local anaesthetic infusion on pain following major lower limb amputation. *Scott Med J* 2008;53(1):4–6.
67. Lambert AW, Dashfield AK, Cosgrove C, et al. Randomized prospective study comparing preoperative epidural and intraoperative perineural analgesia for the prevention of postoperative stump and phantom limb pain following major amputation. *Reg Anesth Pain Med* 2001;26(4):316–321.
68. Sherman RA. A survey of current phantom limb treatments in the United States. *Pain* 1980;8:85.
69. Carabelli R, Kellerman WC. Phantom pain: relief by application of TENS to the contralateral extremity. *Arch Phys Med Rehabil* 1985;66:466.
70. Rusy LM, Troshynski TJ, Weisman SJ. Gabapentin in phantom limb pain management in children and young adults: report of seven cases. *J Pain Symptom Manage* 2001;21(1):78–82.
71. Choi IH, Kumar SJ, Bowen JR. Amputation or limb-lengthening for partial or total absence of the fibula. *J Bone Joint Surg Am* 1990;72:1391.
72. Coventry MB, Johnson EWJ. Congenital absence of the fibula. *J Bone Joint Surg Am* 1952;34:941.
73. Pappas AM. Congenital abnormalities of the femur and related lower extremity malformations: classification and treatment. *J Pediatr Orthop* 1983;3:45.
74. Maffulli N, Fixsen JA. Fibular hypoplasia with absent lateral rays of the foot. *J Bone Joint Surg Br* 1991;73:1002.
75. Maffulli N, Fixsen JA. Management of forme fruste fibular hemimelia. *J Pediatr Orthop B* 1996;5:17.
76. Birch JG, Lincoln TL, Mack PW. Functional classification of fibular deficiency. In: Herring JA, Birch JG, eds. *The child with a limb deficiency*. Rosemont, IL: American Academy of Orthopaedic Surgeons, 1998:161–170.
77. Rogala EJ, Wynne-Davies R, Littlefohn A, et al. Congenital limb anomalies: frequency and aetiological factors. *J Med Genet* 1974;11:221.
78. Baek GH, Kim JK, Chung MS, et al. Terminal hemimelia of the lower extremity: absent lateral ray and a normal fibula. *Int Orthop* 2008;32(2):263–267.
79. Searle CP, Hildebrand RK, Lester EL, et al. Findings of fibular hemimelia syndrome with radiographically normal fibulae. *J Pediatr Orthop B* 2004;13(3):184–188.
80. Bohne WH, Root L. Hypoplasia of the fibula. *Clin Orthop* 1977;125:107.
81. Kalamchi A. *Congenital lower limb deficiencies*. New York, NY: Springer-Verlag, 1989.
82. Boakes JL, Stevens PM, Moseley RF. Treatment of genu valgus deformity in congenital absence of the fibula. *J Pediatr Orthop* 1991;11:721.
83. Pappas AM, Miller JT. Congenital ball-and-socket ankle joints and related lower-extremity malformations. *J Bone Joint Surg Am* 1982;64:672.
84. Takakura Y, Tamai S, Masuhara K. Genesis of the ball-and-socket ankle. *J Bone Joint Surg Br* 1986;68:834.
85. Grogan DP, Holt GR, Ogden JA. Talocalcaneal coalition in patients who have fibular hemimelia or proximal femoral focal deficiency. A comparison of the radiographic and pathological findings. *J Bone Joint Surg Am* 1994;76:1363.
86. Manner HM, Radler C, Ganger R, et al. Dysplasia of the cruciate ligaments: radiographic assessment and classification. *J Bone Joint Surg Am* 2006;88(1):130–137.
87. Beaty JH. Congenital malformations of the lower extremity. In: *Campbell's operative orthopedics*. St. Louis, MO: Mosby-Year Book, 1998: 972–975.
88. Fulp T, Davids JR, Meyer LC, et al. Longitudinal deficiency of the fibula. Operative treatment. *J Bone Joint Surg Am* 1996;78(5):674–682.
89. Herring JA, Barnhill B, Gaffney C. Syme amputation; an evaluation of the physical and psychological function in young patients. *J Bone Joint Surg Am* 1986;68:573.
90. Davidson WH, Bohne WH. The Syme amputation in children. *J Bone Joint Surg Am* 1975;57:905.
91. Thompson TC, Straub LR, Arnold WD. Congenital absence of the fibula. *J Bone Joint Surg Am* 1957;39:1229.
92. Farmer AW, Laurin CA. Congenital absence of the fibula. *J Bone Joint Surg Am* 1960;42:1.
93. Kruger LM, Talbot RD. Amputation and prosthesis as definitive treatment in congenital absence of the fibular. *J Bone Joint Surg Am* 1961;43:625.
94. Wood WL, Zlotsky N, Westin GW. Congenital absence of the fibula: treatment by Syme amputation—indications and technique. *J Bone Joint Surg Am* 1965;47:1159.
95. Hootnick D, Boyd NA, Fixsen JA, et al. The natural history and management of congenital short tibia with dysplasia or absence of the fibula. *J Bone Joint Surg Br* 1977;59:267.
96. Miller LS, Bell DF. Management of congenital fibular deficiency by Ilizarov technique. *J Pediatr Orthop* 1992;12:651.
97. Stanitski DF, Shahcheraghi H, Nicker DA, et al. Results of tibial lengthening with the Ilizarov technique. *J Pediatr Orthop* 1996;16:168.
98. Catagni MA, Guerreschi F. Management of fibular hemimelia using the Ilizarov method. In: Herring JA, Birch JG, eds. *The child with a limb deficiency*. Rosemont, IL: American Academy of Orthopaedic Surgeons, 1998:179–194.
99. Catagni MA. Management of fibular hemimelia using the Ilizarov method. *Instr Course Lect* 1992;41:431.
100. Gibbons PJ, Bradish CF. Fibular hemimelia: a preliminary report on management of the severe abnormality. *J Pediatr Orthop B* 1996;5:20.
101. Ghoneem HG, Wright JG, Cole WG, et al. The Ilizarov method for correction of complex deformities: psychological and functional outcomes. *J Bone Joint Surg Am* 1996;78:1480.
102. Price CT. Editorial: are we there yet? Management of limb-length inequality. *J Pediatr Orthop* 1996;16:141.
103. Naudie D, Hamdy RC, Fassier F, et al. Management of fibular hemimelia: amputation or limb lengthening [see comments]. *J Bone Joint Surg Br* 1997;79:58.
104. Correll J. Management of fibular hemimelia [letter; comment]. *J Bone Joint Surg Br* 1997;79:1040.
105. McCarthy JJ, Glancy GL, Chang FM, et al. Fibular hemimelia; comparison of outcome measurements after amputation and lengthening. *J Bone Joint Surg Am* 2000;82:1732.
106. Herring JA. Symes amputation for fibular hemimelia: a second look in the Ilizarov era. *Instr Course Lect* 1992;41:435.

107. Birch JG, Walsh SJ, Small JM, et al. Syme amputation for the treatment of fibular deficiency. *J Bone Joint Surg Am* 1999;81:1511.
108. Syme J. Amputation at the ankle joint. *Lond Edinb Monthly J Med Sci* 1843;3:93.
109. Anderson L, Westin GW, Oppenheim WL. Syme amputation in children: indications, results, and long-term follow-up. *J Pediatr Orthop* 1984;4:550.
110. Eilert RE, Jayakumar SS. Boyd and Syme ankle amputations in children. *J Bone Joint Surg Am* 1976;58:1139.
111. Fergusson CM, Morrison JD, Kenwright J. Leg-length inequality in children treated by Syme's amputation. *J Bone Joint Surg Br* 1987; 69:433.
112. Mazet R Jr. Syme's amputation. A follow-up study of fifty-one adults and thirty-two children. *J Bone Joint Surg Am* 1968;50:1549.
113. Gaine WJ, McCreath SW. Syme's amputation revisited: a review of 46 cases. *J Bone Joint Surg Br* 1996;78:461.
114. Green WB, Cary JM. Partial foot amputations in children. *J Bone Joint Surg Am* 1982;64:438.
115. Boyd HB. Amputation of the foot with calcaneotibial arthrodesis. *J Bone Joint Surg* 1939;21:997.
116. Hornby RRH. Syme's amputation: follow-up study of weight-bearing in sixty-eight patients. *J Bone Joint Surg Am* 1975;57:346
117. Mazet RJ. Syme's amputation: a follow-up study of fifty-one adults and thirty-two children. *J Bone Joint Surg Am* 1968;50:1549.
118. Serafin J. A new operation for congenital absence of the fibula: preliminary report. *J Bone Joint Surg Br* 1967;49:59.
119. Thomas IH, Williams PF. The Gruca operation for congenital absence of the fibula. *J Bone Joint Surg Br* 1987;69:587.
120. Weber M, Siebert CH, Goost H, et al. Malleolus externus plasty for joint reconstruction in fibular aplasia: preliminary report of a new technique. *J Pediatr Orthop B* 2002;11(3):265–273.
121. Belthur MV, Paley D, Jindal G, et al. Tibial lengthening: extraarticular calcaneotibial screw to prevent ankle equinus. *Clin Orthop Relat Res* 2008;466(12):3003–3010.
122. Catagni MA, Bolano L, Cattaneo R. Management of fibular hemimelia using the Ilizarov method. *Orthop Clin North Am* 1991;22(4):715–722.
123. Cheng JC, Cheung KW, Ng BK. Severe progressive deformities after limb lengthening in type-II fibular hemimelia. *J Bone Joint Surg Br* 1998;80(5):772–776.
124. Kalamchi A, Dawe RV. Congenital deficiency of the tibia. *J Bone Joint Surg Br* 1985;67:581.
125. Loder RT, Herring JA. Fibular transfer for congenital absence of the tibia: a reassessment. *J Pediatr Orthop* 1987;7:8.
126. Fernandez-Palazzi F, Bendahan J, Rivas S. Congenital deficiency of the tibia: a report on 22 cases *J Pediatr Orthop B* 1998;7:298.
127. Goldberg MJ. *The dysmorphic child: An orthopaedic perspective*. New York, NY: Raven Press, 1987.
128. Meyers TH. Congenital absence of the tibia: transplantation of head of fibula: arthrodesis at the ankle joint. *Am J Orthop Surg* 1910;3:72.
129. Babbs C, Heller R, Everman DB, et al. A new locus for split hand/foot malformation with long bone deficiency (SHFLD) at 2q14.2 identified from a chromosome translocation. *Hum Genet* 2007;122(2): 191–199.
130. Schoenecker PL. Tibial deficiency. In: Herring JA, Birch JG, eds. *The child with a limb deficiency*. Rosemont, IL: American Academy of Orthopaedic Surgeons, 1998:209–218.
131. Setoguchi Y. Medical conditions associated with congenital limb deficiencies. In: Herring JA, Birch JG, eds. *The child with a limb deficiency*. Rosemont, IL: American Academy of Orthopaedic Surgeons, 1998:51–60.
132. Schoenecker PL, Capelli AM, Millar EA. Congenital longitudinal deficiency of the tibia. *J Bone Joint Surg Am* 1989;71:278.
133. Aitken GW. Tibial hemimelia. In: Aitken GT, ed. *Selected lower limb anomalies: Surgical and prosthetic management*. Washington, DC: National Academy of Sciences, 1971:1.
134. Wilson GN. Heritable limb deficiencies. In: Herring JA, Birch JG, eds. *The child with a limb deficiency*. Rosemont, IL: American Academy of Orthopaedic Surgeons, 1998:39–50.
135. Grissom LE, Harcke HT, Kumar SJ. Sonography in the management of tibial hemimelia. *Clin Orthop* 1990;251:266.
136. Laor T, Jaramillo D, Hoffer FA, et al. MR imaging in congenital lower limb deformities. *Pediatr Radiol* 1996;26(6):381–387.
137. Lejman T, Sułko J, Lorkowski J. Intraoperative sectional anatomy of congenital absence of tibia type I A. *Chir Narzadow Ruchu Ortop Pol* 1999;64(1):45–51.
138. Sulamaa M, Ryoppy S. Congenital absence of the tibia. *Acta Orthop Scand* 1963;33:262.
139. Brown FW. Construction of a knee joint in congenital total absence of the tibia (paraxial hemimelia tibia). *J Bone Joint Surg Am* 1965;47:695.
140. Christini D, Levy EJ, Facanha FA, et al. Fibular transfer for congenital absence of the tibia. *J Pediatr Orthop* 1993;13:378.
141. Epps CH Jr, Tooms RE, Edholm CD, et al. Failure of centralization of the fibula for congenital longitudinal deficiency of the tibia. *J Bone Joint Surg Am* 1991;73:858.
142. Simmons ED Jr, Ginsburg GM, Hall JE. Brown's procedure for congenital absence of the tibia revisited. *J Pediatr Orthop* 1996;16:85.
143. Wehbe MA, Weinstein SL, Ponseti IV. Tibial agenesis. *J Pediatr Orthop* 1981;1:395.
144. Jayakumar SS, Eilert RE. Fibular transfer for congenital absence of the tibia. *Clin Orthop Relat Res* 1979;139:97.
145. Loder RT. Fibular transfer for congenital absence of the tibia (Brown procedure). In: Herring JA, Birch JG, eds. *The child with a limb deficiency*. Rosemont, IL: American Academy of Orthopaedic Surgeons, 1998:223–230.
146. Jones D, Barnes J, Lloyd-Roberts GC. Congenital aplasia and dysplasia of the tibia with intact fibula. Classification and management. *J Bone Joint Surg Br* 1978;60:31.
147. Wada A, Fujii T, Takamura K, et al. Limb salvage treatment for congenital deficiency of the tibia. *J Pediatr Orthop* 2006;26(2):226–232.
148. Kumar A, Kruger LM. Fibular dimelia with deficiency of the tibia. *J Pediatr Orthop* 1993;13:203.
149. Garbarino JL, Clancy M, Harcke HT, et al. Congenital diastasis of the inferior tibiofibular joint: a review of the literature and report of two cases. *J Pediatr Orthop* 1985;5:225.
150. Pattinson RC, Fixsen JA. Management and outcome in tibial dysplasia [see comments]. *J Bone Joint Surg Br* 1992;74:893.
151. Bose K. Congenital diastasis of the inferior tibiofibular joint. *J Bone Joint Surg Am* 1976;58:886.
152. Tuli SM, Varma BP. Congenital diastasis of the tibiofibular mortise. *J Bone Joint Surg Br* 1972;54:346.
153. Tokmakova K, et al. Congenital deficiency of the tibia. *J Pediatr Orthop* 2003;23:649.
154. Sedgwick WG, Schoenecker PL. Congenital diastasis of the ankle joint. *J Bone Joint Surg Am* 1982;64:450.
155. Giavedoni B, Berry CO. An early leg up. *Orthokinetic Rev* 2003;3:32.
156. Choi IH, Lipton GE, Mackenzie W, et al. Wedge-shaped distal tibial epiphysis in the pathogenesis of equinovalgus deformity of the foot and ankle in tibial lengthening for fibular hemimelia. *J Pediatr Orthop* 2000;20(4):428–436.
157. Wilk B, Karol L, Halliday S. Characterizing gait in young children with a prosthetic knee joint. *Phys Ther Products* 1999;10:20.
158. Aitken GT. Proximal femoral deficiency. *A symposium on proximal femoral focal deficiency—a congenital anomaly*. Washington, DC: National Academy of Sciences, 1969:1.
159. Gillespie R, Torode IP. Classification and management of congenital abnormalities of the femur. *J Bone Joint Surg Br* 1983;65:557.
160. Gillespie R. Classification of congenital abnormalities of the femur. In: Herring JA, Birch JG, eds. *The child with a limb deficiency*. Rosemont, IL: American Academy of Orthopaedic Surgeons, 1998:63–72.
161. Kalamchi A, Cowell HR, Kim KI. Congenital deficiency of the femur. *J Pediatr Orthop* 1985;5:129.
162. Fixsen JA, Lloyd-Roberts GC. The natural history and early treatment of proximal femoral focal dysplasia. *J Bone Joint Surg Br* 1974; 56:86.

163. Goddard NJ, Hashemi-Nejad A, Fixsen JA. Natural history and treatment of instability of the hip in proximal femoral focal deficiency. *J Pediatr Orthop B* 1995;4(2):145–149.
164. Sabharwal S, Paley D, Bhave A, et al. Growth patterns after lengthening of congenitally short lower limbs in young children. *J Pediatr Orthop* 2000;20(2):137–145.
165. Wolfgang GL. Complex congenital anomalies of the lower extremities: femoral bifurcation, tibial hemimelia, and diastasis of the ankle. Case report and review of the literature. *J Bone Joint Surg Am* 1984;66:453.
166. van de Kamp JM, van der Smagt JJ, Bos CF, et al. Bifurcation of the femur with tibial agenesis and additional anomalies. *Am J Med Genet A* 2005;138(1):45–50.
167. Bevan-Thomas WH, Millar EA. A review of proximal focal deficiencies. *J Bone Joint Surg Am* 1967;49:1376–1388.
168. Maldjian C, Patel TY, Klein RM, et al. Efficacy of MRI in classifying proximal focal femoral deficiency. *Skeletal Radiol* 2007;36(3):215–220.
169. Pirani S, Beauchamp RD, Li D, et al. Soft tissue anatomy of proximal femoral focal deficiency. *J Pediatr Orthop* 1991;11(5):563–570.
170. Kant P, Koh SH, Neumann V, et al. Treatment of longitudinal deficiency affecting the femur: comparing patient mobility and satisfaction outcomes of Syme amputation against extension prosthesis. *J Pediatr Orthop* 2003;23(2):236–242.
171. Epps CH Jr. Proximal femoral focal deficiency. *J Bone Joint Surg Am* 1983;65:867.
172. Tsirikos AI, Bowen JR. Patellofemoral arthritis in a fused knee with proximal femoral focal deficiency. *J Pediatr Orthop* 2003;23(5): 643–648.
173. Borggreve J. Kniegelenksersatz durch das in der Beinlangsachse urn 1800 gedrehtd Fussgelenk. *Arch Orthop Unfall Chir* 1930;28:175.
174. van Nes CP. Rotation-plasty for congenital defects of the femur: making use of the ankle of the shortened limb to control the knee joint of a prosthesis. *J Bone Joint Surg Br* 1950;32:12.
175. Kostuik JP, Gillespie R, Hall JE, et al. Van Nes rotational osteotomy for treatment of proximal femoral focal deficiency and congenital short femur. *J Bone Joint Surg Am* 1975;57:1039.
176. Torode IP, Gillespie R. Rotationplasty of the lower limb for congenital defect of the femur. *J Bone Joint Surg Br* 1983;65:569.
177. Friscia DA, Moseley CF, Oppenheim WL. Rotational osteotomy for proximal femoral focal deficiency. *J Bone Joint Surg Am* 1989; 71:1386.
178. Alman BA, Krajbich JI, Hubbard S. Proximal femoral focal deficiency: results of rotationplasy and Syme amputation. *J Bone Joint Surg Am* 1995;77:1876.
179. Veenstra KM, Sprangers MAG, van der Eyen JW, et al. Quality of life in survivors with a van Ness-Borggreve rotationplasty after bone tumour resection. *J Surg Oncol* 2000;73:192.
180. Rodl RW, Pohlmann U, Gosheer G, et al. Rotationplasty: quality of life after 10 years in 22 patients. *Acta Orthop Scand* 2002;73:85.
181. Hillmann A, Hoffmann C, Gosheger G, et al. Malignant tumor of the distal part of the femur of the proximal part of the tibia: endoprosthetic replacement of rotationplasty. *J Bone Joint Surg Am* 1999;81:462.
182. Murray PM, Jacobs PA, Gore DR, et al. Functional performance after tibial rotationplasty. *J Bone Joint Surg Am* 1985;67:392.
183. Steenhoff RM, Daanen HAM, Taminiau HM. Functional analysis of patients who have had a modified van Nes Rotationplasty. *J Bone Joint Surg Am* 1993;75:1451.
184. Fowler E, Zernicke R, Setoguchi Y, et al., Energy expenditure during walking by children who have proximal femoral focal deficiency. *J Bone Joint Surg Am* 1996;78:1857.
185. Westin GW, Gunderson FO. Proximal femoral focal deficiency—a review of treatment experiences. In: Aitken GT, ed. *Proximal femoral focal deficiency: A congenital anomaly*. Washington, DC: American Academy of Sciences, 1969:100.
186. Koman LA, Meyer LC, Warren FH. Proximal femoral focal deficiency: a 50-year experience. *Dev Med Child Neurol* 1982;24:344.
187. Panting AL, Williams PF. Proximal femoral focal deficiency. *J Bone Joint Surg Br* 1978;60:46.
188. Dora C, Bühler M, Stover MD, et al. Morphologic characteristics of acetabular dysplasia in proximal femoral focal deficiency. *J Pediatr Orthop B* 2004;13(2):81–87.
189. Tonnis D, Stanitski DF. Early conservative and operative treatment to gain early normal growth in proximal femoral focal deficiency. *J Pediatr Orthop B* 1997;6:59.
190. Steel HH, Lin PS, Betz RR, et al. Iliofemoral fusion for proximal femoral focal deficiency. *J Bone Joint Surg Am* 2001;83:78.
191. Brown KL. Resection, rotationplasty, and femoropelvic arthrodesis in severe congenital femoral deficiency. A report of the surgical technique and three cases. *J Bone Joint Surg Am* 2001;83A:78–85.
192. McCollough NC. Non-standard prosthetic applications for juvenile amputees. *Interclin Inform Bull* 1963;2:7.
193. Kruger L. Lower limb deficiencies/surgical management. In: Bowker JH, Michael JW, eds. *Atlas of limb prosthetics: surgical, prosthetic, and rehabilitation principles*. St. Louis, MO: Mosby, 1992:759.
194. Bochmann D. Prostheses for the limb-deficient child. In: Kostuik J, Gillespie ER, eds. *Amputation surgery and rehabilitation: The Toronto experience*. New York, NY: Churchill Livingstone, 1981:293.
195. James MA, Bagley AM, Brasington K, et al. Impact of prostheses on function and quality of life for children with unilateral congenital below-the-elbow deficiency. *J Bone Joint Surg Am* 2006;88(11):2356–2365.
196. Clark MW, Atkins DJ, Hubbard SA, et al. Prosthetic devices for children with bilateral upper limb deficiencies: when and if, pros and cons. In: Herring JA, Birch JG, eds. *The child with a limb deficiency*. Rosemont, IL: American Academy of Orthopaedic Surgeons, 1998:397–404.
197. Marquardt E. The multiple limb deficient child. In: Bowker JH, Michael JW, eds. *Atlas of limb prosthetics: Surgical, prosthetic, and rehabilitation principles*. St. Louis, MO: Mosby, 1992:839.
198. Scotland T, Galway H. A long-term review of children with congenital and acquired upper limb deficiency. *J Bone Joint Surg Br* 1983;65:346.
199. Brooks M, Shaperman J. Infant prosthetic fitting: a study of results. *Am J Occup Ther* 1965;19:329.
200. Sorbye R. Upper extremity amputees: Swedish experiences concerning children. In: Atkins DJ, Meier RHI, eds. *Comprehensive management of the upper-limb amputee*. Berlin, Germany: Springer-Verlag, 1989:235.
201. Hubbard SA, Kurtz I, Heim W, et al. Powered prosthetic intervention in upper extremity deficiency. In: Herring JA, Birch JG, eds. *The child with a limb deficiency*. Rosemont, IL: American Academy of Orthopaedic Surgeons, 1998:417–432-.
202. Patton JG. Developmental approach to pediatric prosthetic evaluation and training. In: Atkins DJ, Meier RHI, eds. *Comprehensive management of the upper limb amputee*. Berlin, Germany: Springer-Verlag, 1991:137.
203. Patton JG. Upper-limb prosthetic components for children and teenagers. In: Atkins DJ, Meier RHI, eds. *Comprehensive management of the upper limb amputee*. Berlin, Germany: Springer-Verlag, 1991:99.
204. Garst RJ. The Krukenberg hand. *J Bone Joint Surg Br* 1991;73(3):385–388.
205. Irmay F, Merzouga B, Vettorel D. The Krukenberg procedure: a surgical option for the treatment of double hand amputees in Sierra Leone. *Lancet* 2000;356(9235):1072–1075.
206. Baumgartner R, Asey C. Indications for the Krukenberg hand based on long-term results. *Z Orthop Ihre Grenzgeb* 1994;132(3):180–184.
207. Buck-Gramcko D. The value of the Krukenberg operation and its indication despite modern prosthesis of the hand. *Z Orthop Ihre Grenzgeb* 1954;85(3):460–484. German.
208. Swanson AB, DeGroot Swanson G. Krukenberg-plasty in young patients. *Rehabilitation (Stuttg)* 1990;29(1):19–24.
209. Loosli-Guignard RM, Verdan C. Krukenberg's operation. Indications and limitations. *Ann Chir Main* 1983;2(2):154–159.
210. Alekberov C, Karatosun V, Baran O, et al. Lengthening of congenital below-elbow amputation stumps by the Ilizarov technique. *J Bone Joint Surg Br* 2000;82(2):239–241.
211. Jasiewicz B, Tesiorowski M, Kacki W, et al. Lengthening of congenital forearm stumps. *J Pediatr Orthop B* 2006;15(3):198–201.
212. Muilenburg A, LeBlanc M. Body-powered upper-limb components. In: Atkins K, Meier RHI, eds. *Comprehensive management of the upper limb amputee*. Berlin, Germany: Springer-Verlag, 1988:28.

213. Scott RN. Biomedical engineering in upper-limb prosthetics. In: Atkins DJ, Meier RHI, eds. *Comprehensive management of the upper limb amputee*. Berlin, Germany: Springer-Verlag, 1988:173.
214. Letts M, Davidson D. Epidemiology and prevention of traumatic amputations in children. In: Herring JA, Birch JG, eds. *The child with a limb deficiency*. Rosemont, IL: American Academy of Orthopaedic Surgeons, 1998:235–252.
215. Trautwein LC, Smith DG, Rivara FP. Pediatric amputation injuries: etiology, cost, and outcome. *J Trauma* 1996;41:831.
216. Quigley MJ. Prosthetic management: overview, methods, and materials. In: Bowker JH, Michael JW, eds. *Atlas of limb prosthetics: Surgical, prosthetic, and rehabilitation principles*. St. Louis, MO: Mosby, 1992:67.
217. Steele A. A survey of clinical CAD/CAM use. *J Prosthet Orthot* 1994;6:42.
218. Michael JW. Reflections on CAD/CAM in prosthetics and orthotics. *J Prosthet Orthot* 1989;1:116.
219. Sutherland DH. *The development of mature walking*. Oxford, UK: MacKeith Press, 1988.
220. Keen M. Early development and attainment of normal mature gait. *J Prosthet Orthot* 1993;5:34.
221. Pritham C. Workshop on teaching material for above-knee variants. *J Prosthet Orthot* 1989;1:50.
222. Gottschalk F, Stills S, McClellan B, et al. Does socket configuration influence the position of the femur in above-knee amputation? *J Prosthet Orthot* 1990;2:94.
223. Michael JW. Prosthetic knee mechanisms. *Phys Med Rehabil State Art Rev* 1994;8:147.
224. Gard S, Childress D, Uellendahl J. The influence of four-bar linkage knees on prosthetic swing-phase floor clearance. *J Prosthet Orthot* 1996;8:34.
225. Michael JW. Options meet varying needs. *Motion* 1997;7:34.
226. Barth D, Schumacher L, Thomas S. Gait analysis and energy cost of below knee amputees wearing six different prosthetic feet. *J Prosthet Orthot* 1991;4:63.
227. Cummings K, Kapp S. Lower limb pediatric prosthetics: general considerations and philosophy. *J Prosthet Orthot* 1991;4:196.
228. Neilson DH, Shurr DG, Golden JC, et al. Comparison of energy cost and gait efficiency during ambulation in below-knee amputees using different prosthetic feet—a preliminary report. *J Prosthet Orthot* 1988;1:24.
229. Letts M, Pyper A. The modified Chopart's amputation. *Clin Orthop* 1990;256:44.
230. Coutler-O'Berry C. Physical therapy management in children with lower extremity limb deficiencies. In: Herring JA, Birch JG, eds. *The child with a limb deficiency*. Rosemont, IL: American Academy of Orthopaedic Surgeons, 1998:319–330.
231. Effgen SK. The educational environment. In: Campbell SK, Vanderlinden DW, Palisano RJ, eds. *Physical therapy for children*, 2nd ed. Philadelphia, PA: WB Saunders, 2000.
232. Radocy B. Upper-limb prosthetic adaptations for sports and recreation. In: Bowker JH, Michael JW, eds. *Atlas of limb prosthetics: Surgical, prosthetic, and rehabilitation principles*. St. Louis, MO: Mosby, 1992:325.
233. Michael JW, Gailey RS, Bowker JH. New developments in recreational prostheses and adaptive devices for the amputee. *Clin Orthop* 1990;256:64.
234. Anderson TF. Aspects of sports and recreation for the child with a limb deficiency. In: Herring JA, Birch JG, eds. *The child with a limb deficiency*. Rosemont, IL: American Academy of Orthopaedic Surgeons, 1998:345–352.
235. Paley D, Standard SC. Treatment congenital femoral deficiency. In: Weisel SW, ed. *Operative techniques in orthopaedic surgery*. Philadelphia, PA: LWW, 2011:1202–1223.