

Recent Concepts in the Treatment of the Limb-Deficient Child¹

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STIMULATED in the United States by the development of 22 child amputee clinics (19) during the past 12 years, and ignited by the catastrophic epidemic of congenitally deformed offspring associated with maternal thalidomide ingestion in Europe and Asia during recent years (5,6,10,11,20,21), the limb-deficient child (2) has captured the interest not only of the physician, embryologist, anatomist, and prosthetist, but also of the organic and physical chemist, radiologist, geneticist, sociologist, statistician, psychologist, and, still waiting in the wings, the expert in forensic medicine. While each field has contributed much, this article will confine itself to the advances found in embryology and teratology, training, prosthetics, and surgery.

EMBRYOLOGY AND TERATOLOGY

It should be recalled that the human embryo displays paired mesodermal ridges paralleling the dorsal ridge at approximately the fourth intrauterine week. By the end of the sixth week ectodermal masses appear at the shoulder and hip areas of these ridges, the so-called *limb buds* (8,9,14) (Fig. 1). In the next two weeks these limb buds develop into upper and lower extremities with human configuration

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and microscopically recognizable muscles, tendons, nerves, bones, ligaments, and joints—even to collateral ligaments in the interphalangeal joints of the fingers—and all before

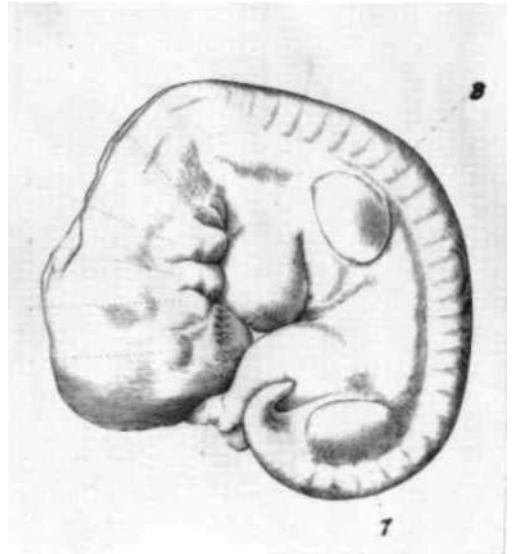


Fig 1. Upper 8 and lower 7 show limb bud development in the six-week human embryo 6.33 mm. in length. From Blechschmidt, E., *The Stages of Human Development Before Birth* (3).

the embryo is more than 25 mm. in length (Fig. 2). The first 56 days of life are devoted to *differentiation* of organs and tissue systems, and the remainder of the 270 days are occupied primarily with growth and development of these tissues. There is increasing evidence that the great bulk of congenital anomalies apparent at birth have occurred during the first eight weeks before the embryo is one inch in crown-



Fig. 2. Development of upper and lower extremities in the eight-week human embryo. Noteworthy is the remarkable growth and differentiation since the limb buds of only two weeks earlier. Patterns of muscles and nerves are comparable to those of an adult. Crown-to-rump length is only 25 mm. From Bardeen, *American Journal of Anatomy*, Vol. 1, 1902.

to-rump length and often before the mother herself is convinced that she is pregnant.

Streeter (18) has defined 22 horizons appearing in the human embryo during the first six weeks. He describes the thickened ectoderm of the upper ridge as Horizon XII, the *Anlage* of the limb buds, appearing at approximately the fourth week along with the paired somites of the dorsal ridge. The somites contribute to the development of vertebral bodies and ribs but probably not, contrary to previous concepts, to the development of limbs themselves. Saunders (15,16,17) has shown that excision of somites results in no alteration of the adjacent limb in the experimental embryo. The ectodermal ridge, however, appears to be the inductor of limb growth. Its removal by microsurgery in the embryos of experimental animals results in the failure of limb development. The transplantation and reorientation of the ecto-

dermal caps of the limb bud will give rise to predicted anomalies. Removal of half the cap results in a limb longitudinally deficient in the area governed by the removed portion. Saunders, Zwilling (22,23), and others have produced twinning, mirror images, and upper-extremity/lower-extremity transplants by such microsurgical methods. The interrelationship of ectoderm and mesoderm is of much interest; one appears to control the growth and differentiation of the other with a distinct specificity of their site of origin. The thickened terminal portion of the limb bud ectoderm is well recognized and is capable of directing and orienting extremity growth.

Blechsmidt's studies (3) of the ectoderm-mesoderm relationship point out the role of cell death or retardation on the concave portion of the limb coupled with cell multiplication on the convex portion as the controlling factor in limb curvature and rotation. Cell death plays a prominent role in the contouring and sculpturing of the embryonic limb to its eventual human form. The latter is evident and specific at the end of the eighth intrauterine week. Selective cellular regression allows separation of mesodermal masses into the parallel skeletal rays of hands and feet, produces the cavitation of joints and articular spaces in the proper locations, and governs the development of body cavities wherever these are located (Fig. 3). Disturbance of either the mechanism of cell reproduction or selective cellular death can be seen as a cause of congenital deformities. The lack of appropriate cell death between femur and tibia will result in a permanently fused knee. The lack of cell death in the interdigital spaces will result in mittenlike syndactylized hands.

The maintenance or guiding factor governing any or all of these phenomena is as yet undiscovered. Heretofore the cell has been the principal object of study; but lately investigation of the ground substance, the vast intercellular ocean, suggests that the controlling agents may reside in this area of embryonic tissue. Pinner has shown by staining methods that the mesenchymal ground substance is high in mucopolysaccharides, particularly in the region of neural tube closure and in the

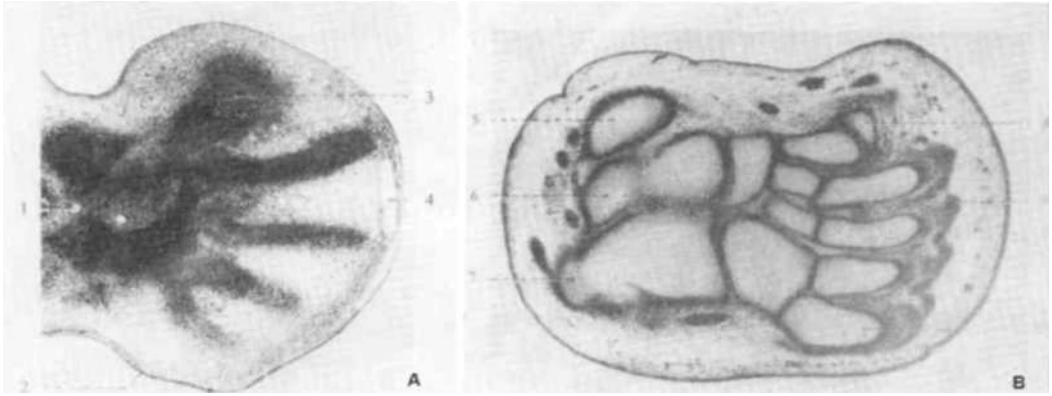


Fig. 3. Selective cellular regression. A, Early cellular regression allowing mesoderm separation into metatarsal rays (14-mm. embryo); B, continued cellular regression producing articular spaces and individual skeletal elements of foot (29-mm. embryo). From Blechschmidt, K., *The Stages of Human Development Before Birth* (3).

basement membrane between the mesoderm and ectoderm of the developing limb bud. Among the substances proven to inhibit mucopolysaccharide biosynthesis are many known teratogenic agents. The embryonic age at which the anomaly production occurs may be as varied as the number of agents which can produce them with regularity in the laboratory animal. Some 70 agents, insults, or mechanisms are known methods that may produce statistically significant deformities. The timing of administration within the fetal growth period seems as important as the nature of the agent itself. The complex pattern of cellular growth and cellular death may be interrupted at any point, producing a characteristic anomaly.

Zwilling points out that deformities may result from: *first*, an interference with *progressive events* of either the primary limb bud establishment or the elaboration of secondary limb patterns; *second*, interference with the *regressive* phase of selective cell death; *third*, interference with the *growth and development* of elements once established; and, *fourth*, interference with the as yet little known molecular control of induction and regression. Molecular control appears to be an enzyme-coenzyme relationship that may be the basis for the first three categories.

The role of thalidomide (Fig. 4) is not yet understood. Its mechanism of action suggests that it may be a counterfeit agent interjecting itself into a vital process but preventing the

successful completion of that physiological process. It appears to have been a definite factor in the recent wave of congenital deformities among maternal populations using the medication during early weeks of pregnancy. Carefully documented case histories as well as thoroughly checked epidemiological studies not only reveal close relationship between the day of starting the drug and the type of anomaly observed at birth, but also have shown the rise in incidence of certain infant deformities within a geographical area to be in direct relationship to the amount of thalidomide supplied to the wholesale drug agents of that particular locale. The critical maternal ingestion period appears to have been between the 27th and 30th day of pregnancy; the most severely deformed children seem to have resulted from medication taken between the 27th and 30th day after conception.

In addition to the environmentally induced deformity, anomalies may also result from chromosomal aberration within the infant itself as well as from genetic potentialities of the parents. Patterns of deformity on an hereditary basis are well recognized. To the parents of deformed children the genetic possibilities are of marked importance: "If we have another baby will it be deformed? If this deformed baby grows up and marries, will it have deformed offspring?" The complexity of eugenic counselling is beyond the scope of this paper, but, in general, the answer to either



Fig. 4. Three-year-old boy whose mother took thalidomide during early pregnancy. He exhibits typical facial and ear deformities plus bilateral upper-extremity amelia with rudimentary single digits. Successful application of externally powered prostheses will give major benefits. Such patients should be trained in the use of their feet in all aspects of daily activities. This patient has won an award at a painting exhibition, competing against older normal children.

question would be "no risk" with environmentally induced deformities (drugs, viruses, X-ray exposure, etc.). With chromosomal aberration the risk is probably small. With a congenital malformation due to dominant or recessive genes in both parents the chances of another deformed offspring are strong, and the chances of this child passing the deformity to his children are also strong. It behooves the clinician to ascertain the basic cause of his patient's difficulty in order to provide parents with this vital information.

Whatever the etiology, deformities can be classified after the method of O'Rahilly (13)

into two basic types—*terminal* where no part exists distal to or in line with the deficient portion, and *intercalary* where there is an intersegmental loss with portions proximal to and distal to remaining grossly intact. Within both of these classifications, the deficiencies may be *transverse*, encompassing the entire girth of the limb; or they may be *paraxial*, occurring as a longitudinal deficiency in grossly the pre- or postaxial elements with the uninvolved portion remaining grossly intact. While no universally accepted explanation of these deformity types has yet been made, the experimental findings of Saunders and Zwilling as previously noted can be considered as a possible explanation of human deformities.

The dermatome distribution of spinal nerves in the fetus reveals a pre- and postaxial distribution of known regularity. By using a diagram of these patterns primarily for the purpose of orientation, the two basic deficiency types, *terminal* and *intercalary*, can be assigned one to each side of the diagram, and the two subtypes, *paraxial* and *transverse*, can be assigned one to an upper and the other to a lower limb (7). With this simple arrangement the skeletal deficiencies of O'Rahilly's classification can be related very closely to the findings of the experimental embryologists and teratologists. The Frantz-O'Rahilly (4) method of classifying clinical entities has proven very useful in the cataloguing and division for treatment of child patients with these congenital deficiencies. This is based on the roentgenographic appearance of the extremities observable within a day or two after birth.

TRAINING (Fig. 5)

Training of the limb-deficient child actually starts within a few hours of birth when the parents are provided with a detailed, factual, realistic, and above all, sympathetic appraisal of their baby and its prospects for future educational, vocational, and social rehabilitation (12). The psychic trauma to the parents should not be underestimated, and a well-founded program can do much to alleviate their burden. The child should become a part of the family immediately. Later, prolonged hospitalization may prove necessary for surgical and prosthetic treatment. The first few months within the



Fig. 5. Training by a competent therapist ensures successful use of a well-fitted prosthesis.

protective love of an informed family appears to be of great value in the care of these patients. The mother will ultimately become the child's best therapist, and the early months of intimate association will provide a basis for her later role as teacher. The mother must be given systematic instruction that is checked and supervised at regular intervals.

Physical and occupational therapy should be started as soon as the youngster begins to take part in his environment. The prone position will strengthen the back as the youngster lifts his head in curiosity. Clothing should be altered so that foreshortened extremities have full freedom to touch and grasp. In the armless child the legs should be completely uncovered so that the baby can watch and play with his feet. These will develop into his future "hands" and toys should be placed for foot activities. Even the weakest and most rudimentary digits arising from malformed shoulders should be stimulated and strengthened for eventual prosthesis control. The ability to grasp and touch must be learned; lack of opportunity to

do this will result in atrophy and disuse of valuable extremities. Children with both upper and lower amelias must be encouraged to feel with their mouth and lips and to hold objects between their chin and shoulders. Flexibility of the legs and feet should be encouraged in the armless. Surgical procedures should aim to increase function, and those that limit the ability of the feet to reach the face should be avoided. Hip flexibility must be much above the normal, but once obtained, it can provide these patients with an independence in dressing and undressing as well as in handling their own toilet care. Walking will be somewhat delayed in the child with foreshortened upper extremities. The ability to pull himself up, to balance when erect, and to cushion himself when falling will be absent. His head should be protected with a sponge-rubber-ring helmet (Fig. 6). He should be deliberately taught to fall and rise without injury.



Fig. 6. Leather-and-sponge-rubber bicycle helmet protects the toddler-age patient with bilateral arm deficiencies against head injuries.

How does one decide that an arm prosthesis is necessary? Probably a child who can eat and play with his deformed hands will not require a prosthesis. The hands should be capable of apposition across the front of the body, even with the expansion of the thorax in growth. Youngsters with amelias or short phocomelias will require artificial limbs, and fitting starts when the child has obtained good sitting balance. Children whose lower-extremity deformity prohibits stabilization in sitting should be provided with appropriate stabilized trunk support for upright posture. The arm prostheses are fitted initially for *passive* operation (Fig. 7)—joints, terminal devices, etc., are operated by the parents, or by the child's remaining good hand in the unilateral amputee. Gross arm movement bringing both hands together in a clapping motion will serve to hold a ball or doll and is actually the first *active* motion of the amputee. This is basically a shrugging or coapting movement of the shoulders. When this is mastered, active control of

one of the terminal devices, usually on the dominant side first, can be added. In the amputee fitted passively at six or eight months, this can be accomplished at the sixteenth to the twenty-fourth month depending on the ability of the child and the nature of his deformity. The control of the elbow lock can be added at three years. In the pneumatically powered devices developed in Germany (Fig. 8), the clever amputee has learned good control of elbow flexion and extension, forearm pronation and supination, and terminal-device grasp and release—all under the control of amelic shoulders or phocomelic fingers—by five years of age. The use of external power, either from portable carbon-dioxide-filled tanks or from electrical batteries, is a realistic solution to the complete lack of upper extremities. At the UCLA Child Amputee Prosthetics Project, it has been possible to provide functioning upper extremities for these patients with harnesses and cables, but the energy expense to the patient is tremendous and is often not available until

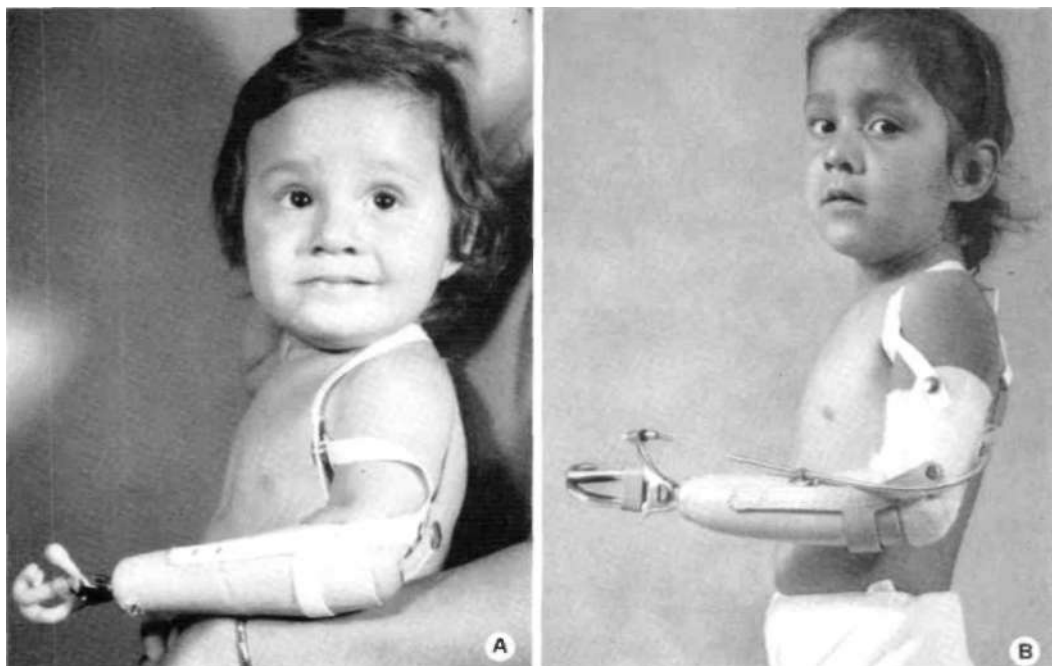


Fig. 7. Upper-extremity prostheses for young patients. A, Passively operated below-elbow prosthesis can be fitted when the child acquires good sitting balance at the age of nine to ten months; B, activation of the terminal device by means of a figure-eight Dacron harness and Bowden cable is possible at 18 months, with good use of the prosthesis in the activities of daily living.

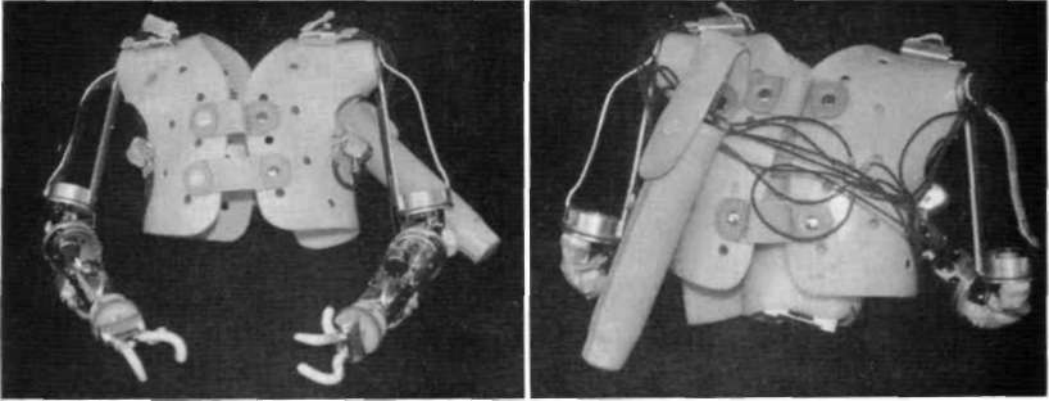


Fig. 8. Anterior and posterior views of pneumatically powered bilateral shoulder-disarticulation prosthesis as constructed under the direction of Dr. Ernst Marquardt, of the University of Heidelberg.

the child is approximately seven or eight years of age.

The training of a child fitted with a prosthesis is a time-consuming but highly rewarding procedure. It is started under the careful supervision of a trained therapist, and the mother is taught as well as the patient. Training then proceeds at home with clinic visits at regular intervals for solution of difficulties and addition of new maneuvers or activities. The functional level of a normal child of the same age should be the basis of achievement goals. A three-year-old with prostheses should not be expected to tie his own shoes when the normal youngster accomplishes this only after diligent instruction at the age of five or six!

The development of foot activities in the armless child should continue all through his life. Prostheses, however good, can never entirely replace well-trained feet. In the bath and in the bed, prostheses are of little help. One patient is a lovely twenty-eight-year-old mother of two active children who drives, keeps house, dresses the children and herself, and fixes her own hair in the current fashion. She is a bilateral upper-extremity amelic and has never worn prostheses, stating that she "can't be bothered." In truth it is considered that she would benefit from artificial arms under many circumstances, but her life has been full without them.

PROSTHETICS

Recent advances in the field of children's prosthetics include improved design and func-

tion of terminal devices (not just "scaled-down" adult hooks), wrist and elbow joints, lightweight plastic sockets and shells, and more efficient harnessing methods. The externally powered, carbon-dioxide devices of Heidelberg, as well as the electrically powered devices of Chicago, are an immense step in the treatment of the bilateral upper short phocomelic and amelic patient. The initial research and development of self-propelled wheeled vehicles for the lower-extremity amputee proceed both in America and Europe (Fig. 9).

The application of prosthesis to the congenital or traumatic child amputee has reached a stage of acceptance in the United States so as to be almost routine. No longer must the child "wait 'til he grows up, then he can get a wooden arm." Earlier and earlier fittings result in complete patient and family acceptance of and benefit from the prosthesis. All patients are fitted with arms at six months of age and with legs at about nine months. The prostheses become as much a part of the youngster as his undergarments and are removed only when the latter are not normally worn—in the bath or in bed. Alterations and refitting accommodate changes inherent in growth and increased activity levels. The capabilities of such patients over their unfitted similarly handicapped counterparts are obvious.

The fitting of the bilateral upper amelic or phocomelic patient has been a very difficult problem. The tremendous rise in the incidence of such patients associated with thalidomide has been seen in many areas. In Schleswig-



Fig. 9. Battery-powered electrical cart. Speed, direction, and seat height selection are controlled by the amputee by means of joystick arrangement. The cart was developed and constructed at the Child Amputee Prosthetics Project, University of California, Los Angeles.

Holstein *three* phocomelias in 266,599 live births were recorded in the 1949-56 period. In September 1961 the rate of *five* phocomelias *per one thousand* births was recorded. Prosthetic restoration of such patients under the classic methods was completely inadequate. Extremities with sufficient length, power, and excursion of movements simply were not available for the successful stabilization and operation of the routine child prosthesis. In Heidelberg, however, a pneumatically powered prosthesis for the adult had been developed. Through clever engineering, painstaking manufacture, and imaginative application, this device was scaled to the infant and is now fitted at about six to eight months of age. The utilization of shoulder motion or phocomelic digit control allows performance of the activities of daily living and play commensurate with the child's age.

Such patients are initially fitted with bilateral shoulder and thorax sockets and taught controlled shoulder motion that brings the prosthetic hands into apposition in a clapping motion. This apposition of the terminal devices in front of the child allows the holding of large balls, dolls, and other toys at a very early age. Release is accomplished by a reversal of shoulder action. These controlled motions produce internal and external rotation of the humeral segments, which in themselves provide the apposition and separation of the hand elements. As maturation and dexterity develop, pneumatic controls for the opening and closing of the terminal hook, for pronating and supinating the forearm, and for the flexion and extension of the elbow can be added in amputees as young as four years. Clever placement of the control valve for phocomelic digit operation or shoulder motion in the amelic, allows a "normal" neuromuscular pattern to develop. For example, the bilateral phocomelic five-year-old opens and closes the hook on his ipsilateral side by digit function; he controls pronation and supination of the forearm—actually wrist rotation—by contralateral digit operation; and he operates flexion and extension of his elbow by ipsilateral operation of a strategically placed valve by the nudge of his chin. The control motions are readily understood by the child; and to see a completely armless six-year-old eat a full meal without assistance, using prostheses that have the gross form of upper extremities and appear very natural beneath clothing, is indeed most gratifying. The problem of carbon-dioxide supply, the maintenance of intricate valves, and the adjustments for growth are readily apparent difficulties. They can be overcome in most instances, however, and the benefits to the patient are obvious.

SURGERY

Orthopaedic surgery in the limb-deficient child includes most of the procedures associated with normal children of the same age, but the frequent association of other organ system anomalies—cardiac, pulmonary, gastrointestinal—requires the close cooperation of experienced pediatricians and anesthesiologists. Routine techniques for monitoring the vital signs of anesthetized patients may become

quite a problem in this group. How does one obtain the blood pressure in an amputee whose limbs end above the elbows and knees? How does one provide parenteral fluids to a quadrilateral phocomelic patient? What are the effects of the all-encompassing surgical drapes on the body temperature of a patient who is deficient in one-half of his normal radiating skin surfaces? How does one secure postoperative dressings and casts on grossly foreshortened extremities? None of these problems are insurmountable, but they do add a complicating factor to what might at first appear as a routine surgical exercise.

In general, one should be overly cautious in the "corrective" surgery of limb deficiencies. Too often the result is an "improved" X-ray picture and a functionally impaired patient. The osteotomies and fusions to realign bizarre malpositions in the upper extremity are a case in point. The arm may appear much straighter after the deviated hand is fused to the single

bone forearm, but the fibrous ankylosis of the elbow, almost always present in these cases, prevents the postoperative patient from reaching his face with his newly aligned hand. He has gained little and lost much. The fusion of unstable knees in an upper-extremity amelic patient may provide a stable gait, but the youngster cannot reach his mouth with his toes—a catastrophe to this type of patient. Ill-conceived amputations of what at first may appear as useless and grotesque appendages frequently remove elements that could mature into useful limbs capable of stabilizing a prosthesis or providing control operations to a power system.

Conversion is a term employed to describe the amputation of an extremity so deformed as to be cosmetically and functionally incapable of developing into a useful member. The limb is *converted* to a satisfactory stump (1). While one should delay ablative surgery in most instances, the life history of certain deformities

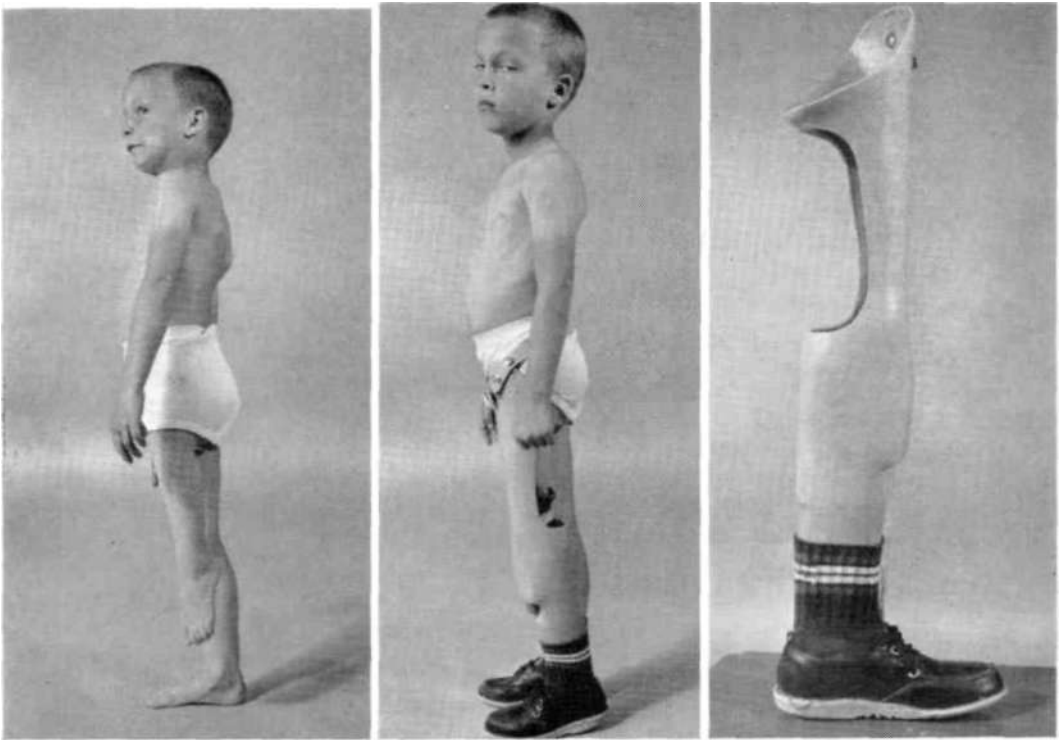


Fig. 10. Views of phocomelia of left lower extremity with pylon-type, end-bearing plastic socket that provides fair cosmesis in long trousers but poor gait characteristics.

is now so well documented that early amputation at the proper level becomes the procedure of choice. Paraxial hemimelias of the lower extremity are common examples. In addition to the complete or partial absence of the tibia or fibula, the remaining bone is shortened, bowed, and rotated; the ankle joint is dislocated; and the foot lies in a plantarflexed attitude of severe valgus or varus. Diligent plaster-of-Paris cast correction, soft tissue surgical releases, and occasional arthrodesis may provide a plantigrade weight-bearing foot. As the child matures, increasing inequality of leg length occurs; additional shoe lifts or braces are added; and by his mid-teens, the patient is walking on a four- to six-inch platform. Or, one might have elected to hold the foot in marked equinus, allowing the tiptoe position to compensate for the decreased leg length.

The bulky prosthesis is then molded about the plantarflexed foot. Such procedures may serve well in the male, whose trousers mask the irregular contour of the prosthesis (Fig. 10). In the female, however, such a prosthesis is impossible to hide or disguise. Functionally, gait patterns are disturbed; and fitting and alignment are complicated. The early conversion of this anomaly by disarticulation of the ankle provides an excellent weight-bearing stump of the Syme type when "barefoot," and the application of a prosthesis provides a cosmetically contoured and finished artificial leg with excellent gait characteristics (Fig. 11). At maturity these stumps have shortened to the conventional below-knee level and are fitted as such.

The congenital reduction in leg length may be so severe that the total length of the af-

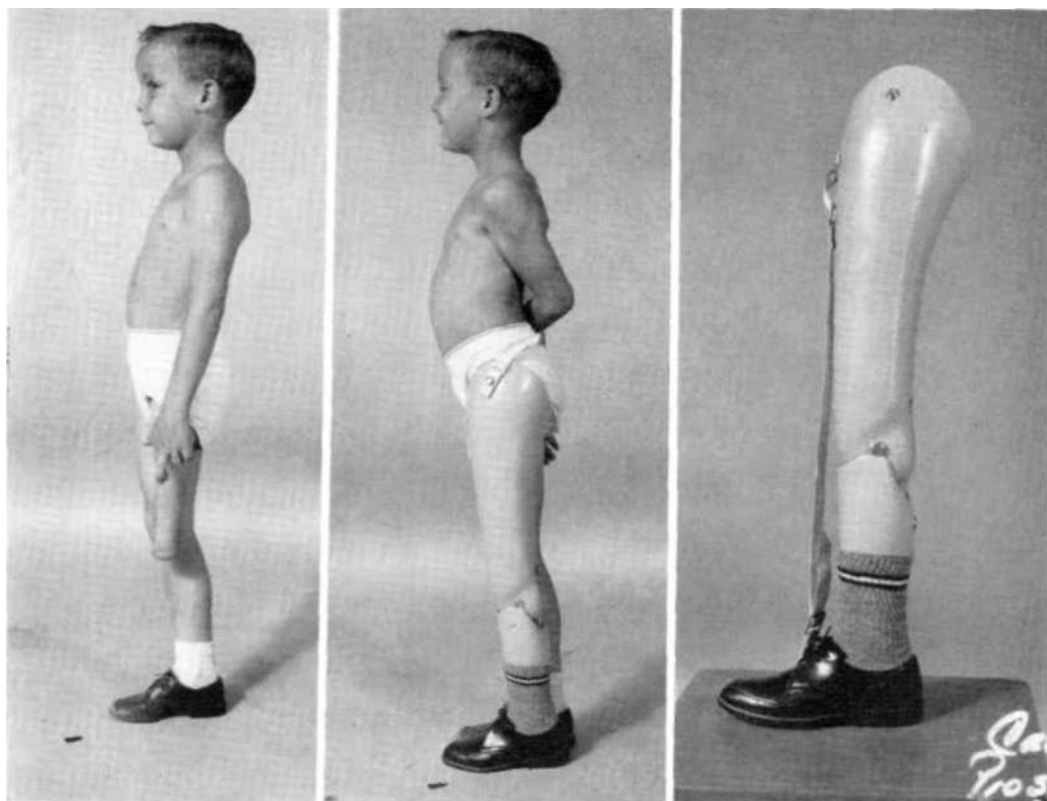


Fig. 11. Same patient who appears in Figure 10 is shown with his prosthesis after a Syme's amputation at the left ankle. End-bearing prosthesis now has knee joint with improved gait characteristics. Unequal future growth will reduce the relative length of the left stump and provide the mature amputee with a stump equal to a long above-knee amputation.

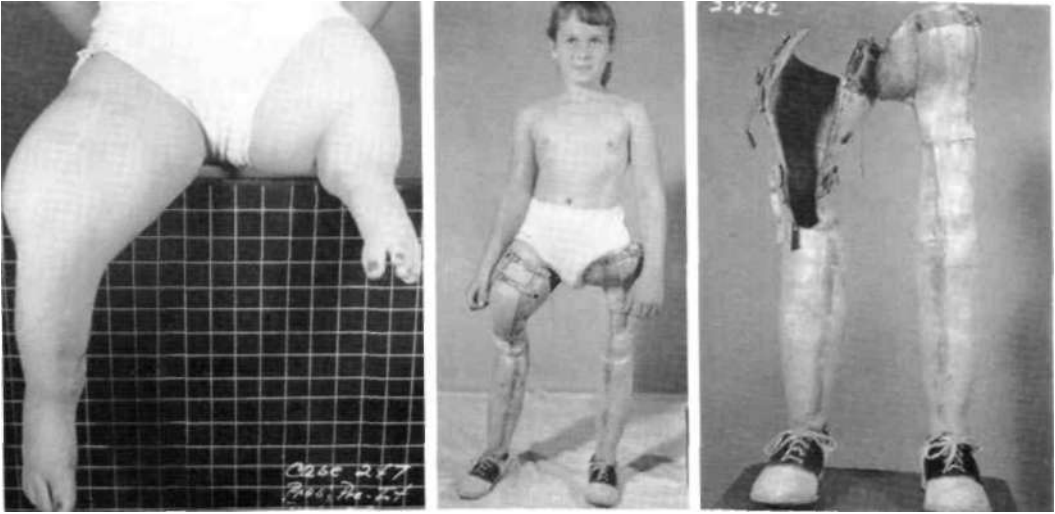


Fig. 12. Views of bilateral phcomelic patient with intact knee and ankle joint on right, fitted with bilateral pylon-type prostheses giving a waddling gait and poor sitting characteristics.



Fig. 13. Same patient shown in Figure 12 after arthrodesis of knee in full extension and Syme's amputation at ankle to provide a stable right thigh stump easily fitted with a standard suction socket. The left phocomelic extremity has been fitted with a modified above-knee socket. Appearance and gait patterns are much improved.

fectured femur and tibia combined may only approximate that of the normal femur. A common multistage procedure, yielding good results at UCLA, consists of an arthrodesis of the knee in full extension, followed, after complete healing, by disarticulation at the ankle joint. A stable, weight-bearing stump of normal "thigh bone" length is provided, and the patient is fitted as a routine above-knee amputee. The gain in function and appearance is very pleasing to the patient and surgeon alike (Figs. 12 and 13).

Fusions in such cases are often performed prior to the closure of epiphyseal plates about the involved joint. Epiphyses fuse readily to their juxta-articular counterpart after excision of joint structures, and their growth contribu-

tion to extremity length continues in spite of operative removal of their articular portions and their transfixion by intramedullary pins used for postoperative stabilization. Arthrodeses in other areas are utilized for stabilizing elements congenitally deficient in supporting ligaments, tendons, and muscles. Transplantation of single remaining bones in paraxial deficiencies of the forearm and leg, with fusions to proximal and distal elements, may provide longer and stronger extremities for use with or without prostheses.

Bizarre abnormalities of the hand may be corrected surgically with much gain in function. The removal of rudimentary elements may provide much increased prehension capability in the remaining structures. *Syndac-*



Fig. 14. Attractive early teen-age female with very short below-elbow congenital amputation fitted with step-up hinge and figure-eight harness. The patient was an excellent wearer but objected to the bulk of the prosthesis and the poor appearance of the shoulder harness. She requested a cineplasty to allow the wearing of strapless dresses.

tylism is a common finding, and the surgical separation of digits is very rewarding. Soft-tissue fusions of terminal phalangeal elements are not rare. Small skin bridges may be painlessly divided by the application of necrotizing surgical silk ligatures. Healing keeps pace with the necrosis, and fingers separated in this fashion often have undetectable scars. Full-length syndactylism will require careful plastic reconstruction at the proper age, using full- and split-thickness flaps and grafts to provide satisfactory noncontracting web space construction as described by Bunnell and others.

Biceps lineplasties have been performed with good results in these patients (Figs. 14, 15, and 16). Below-elbow terminal hemimelias are the commonest major congenital amputation in the UCLA child amputee clinic case load. While there is always hypoplasia of humeral musculature, biceps and brachialis development is sufficient to produce excellent

force and excursion of the cineplasty tunnel as well as stump movement. The removal of the annoying and cumbersome shoulder harness, coupled with the increased power and flexibility of the prosthesis, has made this an excellent procedure in the properly selected case. It is possible that it should be recommended more often for teen-age patients.

The most common single surgical procedure in the UCLA child amputee clinic is stump revision for *terminal skeletal overgrowth* (Fig. 17). For the most part, this annoying phenomenon occurs in the above-elbow and the below-knee stumps—no cases in the congenital forearm or femoral amputees have been observed at the UCLA child amputee clinic, although they have occurred in all locations in the acquired amputee. While previous authors have stated that terminal overgrowth does not occur in the congenital type, it has been found to be so common at the L'CLA child amputee clinic that the parents of these patients are warned at

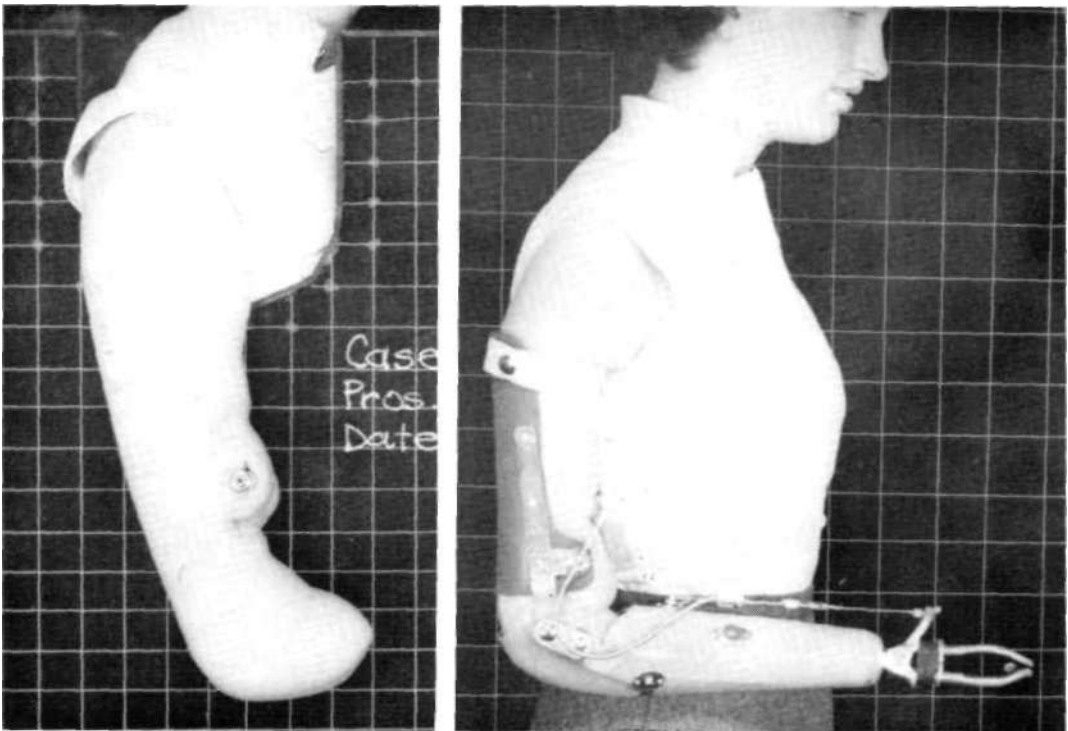


Fig. 15. Same patient shown in Figure 14 after a biceps cineplasty had been performed. The figure-eight harness has been discarded. Hand or hook can be attached interchangeably to the split-socket prosthesis.

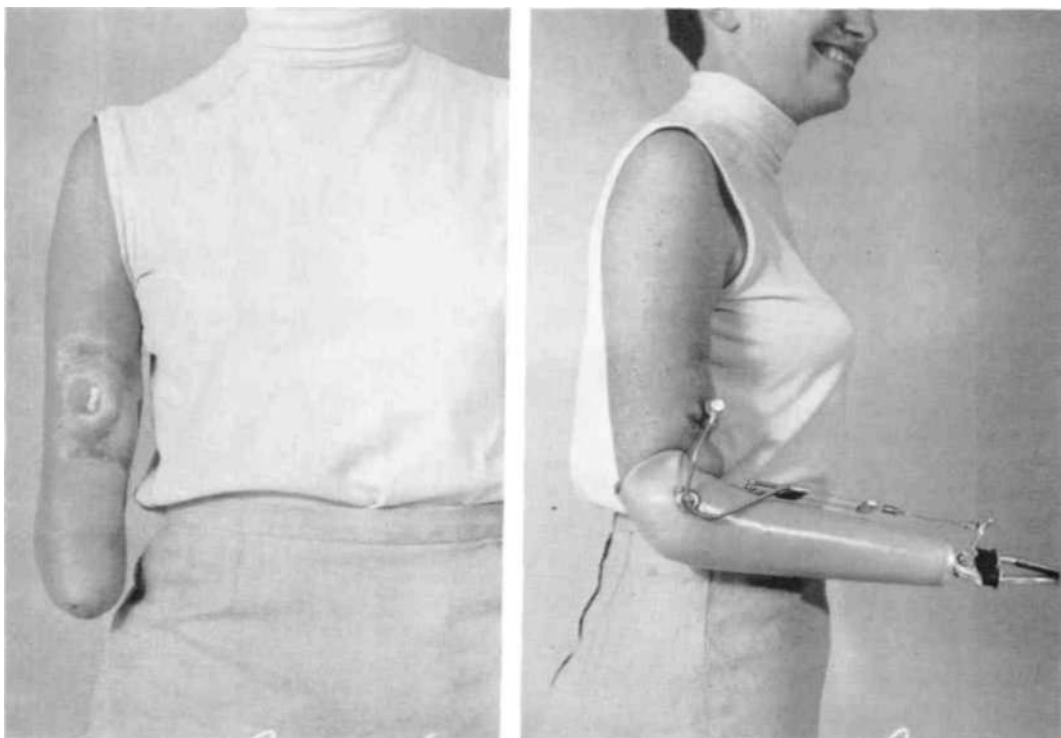


Fig. 16. Same patient shown in Figures 14 and 15 fitted with a Munster-type socket. Cineplasty tunnel is in excellent condition. The patient is a full-time wearer and is delighted with the reduced weight and complexity of the prosthesis as well as with increased proprioception and the stability of the fitting.

their initial clinic visit. Subsequent protrusion of the underlying bone into the terminal stump comes as no surprise, and parents are instructed that one to five surgical revisions may be necessary before the child reaches full growth and the difficulty ceases. Various techniques in the treatment of the bone end have been attempted—including osteoperiosteal plugs, bars, flaps, etc.—to decrease the growth potential of the severed bone end, without success. The transected cortical shaft has been compared to the one side of a long-bone fracture—the terminal appositional subperiosteal and endosteal bone growth is quite similar to the process seen in fracture healing. Why it should produce a sharp, soft-tissue-penetrating, terminal spike is not understood. A bursal sac is almost always present. The routine revision consists of a transverse terminal incision into the sac, removal of the protruding spike with all of its soft tissue covering two to three centi-

meters proximal to the tip, and excision of the proximal bursal sac wall. Complete excision of the entire bursa often complicates distal healing, and since the bursa forms again—whether removed or not—the distal wall is left undisturbed. Healing is usually prompt, and prostheses may be refitted in four to six weeks after surgery.

SUMMARY

In summary, continued research in the fields of embryology, teratology, and genetics has indicated that congenital abnormalities are not always "inherited" but may be environmentally induced, may be caused by chromosomal aberrations, or may result from genetic backgrounds. In a high percentage of cases, the defect has occurred by the end of the eighth intrauterine week in human embryos. The exact mechanism of action of teratogenic agents is not completely understood; but,

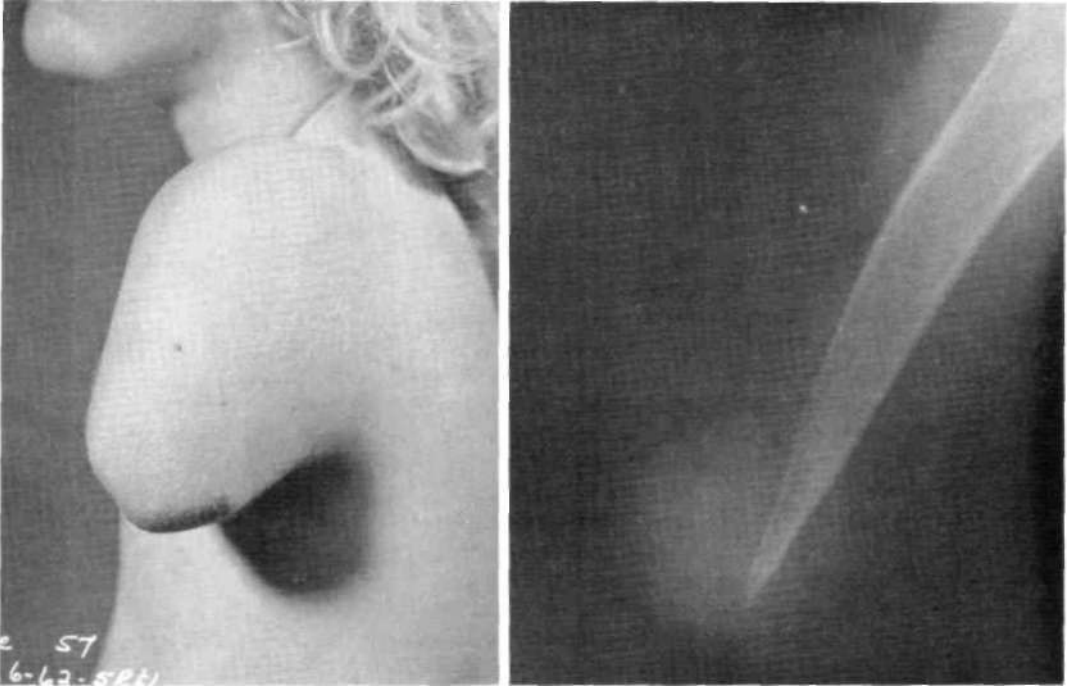


Fig. 17. Congenital above-elbow amputee with recurrent episodes of terminal bone overgrowth and stump perforation. X-ray reveals sharp spur formed by terminal appositional new bone formation. Frequent surgical intervention is required in humeral and tibial amputations of this type; revision is seldom necessary in forearm and femoral amputations.

experimentally, the embryos of laboratory animals can be insulted by a number of agents producing predictable deformities. The time of the insult appears as important as the nature of the agent itself.

Based on roentgenographic findings in the newborn, congenital deformities can be classified according to the methods of O'Rahilly and Frantz, allowing programs and patterns of rehabilitation on an organized basis.

The early training of limb-deficient children allows them to capitalize on existing extremities and simplifies eventual prosthetic restitution.

Advance within the prosthetics field results in the present practical use of externally powered devices in the child amputee and predicts continued developmental success in the future.

In general, surgery should be deferred until maturation produces the maximum potential in the involved limb. The life history of certain deformities, however, allows conversion to

functional amputation stumps at an early age; and the subsequent early fit with prosthetic devices greatly enhances the patients' habilitation.

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