"Pylon-Prosthetic" Devices for Lower Extremity Congenital Skeletal Limb Deficiencies

(From the Shriner's Hospital for Crippled Children, Minneapolis, Minnesota)

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Preface

Rapid advances in prosthetics since World War II have made it possible to successfully fit amputees of all types, including those with stump deformities, with much greater ease and better results than at any previous time. Improved fitting techniques, materials, and many improved mechanical components now available to the prosthetist provide the amputee of today a much better device with improved functional capabilities, comfort and cosmesis.

These improving techniques of the past twenty years more recently have been applied to the child amputee with ever increasing success despite the problems that are presented in the care of children, particularly those with congenital skeletal limb deficiencies. The success in fitting of the so-called standard child amputee has stimulated interest in attempting to provide better bracing and prosthetic substitutions for children with the more bizarre deficiencies.

Nomenclature and Embryology

The various deficiencies present a frustrating problem to the surgeon and prosthetist-orthotist as well. Communications with respect to prosthetic or brace fitting of the more bizarre limb deficiencies has in the past been very limited because of the lack of a common language and nomenclature. Five years ago youngsters with such limb deficiencies had lobster-claw deformities, short legs, flippers, etc. The so-called lobster-claw deformity had as many as ten synonyms, including crayfish-claw pinchers, adactyly, etc. These synonyms were used to describe partial adactylia.

During the past fifteen years the early development of limbs in the human embryo has been investigated in detail by many. The limb buds appear on the lateral body wall at four post ovulatory weeks. The buds grow and differentiate rapidly within the ensuing three weeks, developing in a proximal to distal sequence. The skeletal elements of the limb buds are first mesenchyme condensations which rapidly chondrify in sequential order, followed by ossification. Ossification begins at five post ovulatory weeks in the clavicle and seven post ovulatory weeks in the long bones. Initially, diaphysis bone collar formation develops rapidly, followed by enchondral ossification. By seven post ovulatory weeks, the skeletal elements of the limb with the exception of the clavicle are individually present in detail as cartilagenous models, as are the extremity joints, with the larger joints beginning to show cavitation.

In summary, the skeleton is, for practical purposes, well developed early as the miniature of post-natal life. Congenital skeletal limb deficiencies therefore arise during the first seven weeks of intrauterine life or during the embryonic phase of intrauterine life. For the remainder of intrauterine life, recognized as the fetal period, development is growth rather than differentiation.

Congenital skeletal limb deficiencies are now classified by descriptive terms derived from the Greek language. The three basic terms are *amelia,* absence of a limb; *hemimelia,* absence of half of a limb; and *phocomelia,* flipper type of limb where the hand or foot are attached more proximally to the trunk. *Acheiria* and *apodia* are the terms used for absence of the hand and foot. *Adactylia* is absence of a digit, usually associated with absence of the proximal metacarpal or metatarsal bone. *Aphalangia* is the term used for absence of phalanges. Hemimelia may be complete or incomplete, the former when the distal half of the limb is absent and the latter when a greater portion of the distal half is absent. *Paraxial hemimelia* refers to axial deficiency of an extremity, absence of a radius or ulna or tibia or fibula. To localize the skeletal deficiency, the terms *terminal* and *intercalary* are now used, the former when parts are absent distal to or in line with a deficient or absent portion and the latter where the mid portion of the limb is missing, the proximal and distal portions relatively normal. Each of these types may be transverse, the deficiency extending completely across the limb or longitudinal, a long axis deficiency or absence. Phocomelia, complete or incomplete, may be proximal or distal. So-called proximal femoral focal deficiency can be classified as incomplete proximal phocomelia. Complete phocomelia is uncommon as usually some vestige form of ossification sooner or later develops within the absent skeletal site.

The use of this classification should serve as a source of common language and communication for the surgeon and the prosthetist-orthotist.

Description and Objectives

The purpose of this discussion is the application of some of the improved fitting techniques, materials and mechanical components in providing improved "Pylon-Prosthetic" devices for some of the more bizarre lower extremity congenital skeletal limb deficiencies. Heretofore, children with deficiencies other than amputations and not severe enough to require surgical modification to amputations, generally were fitted with some type of pylon, usually the basic article, a crutch or a modified shoe. For those deficiencies such as proximal complete or partial femoral phocomelia, the youngster usually was provided with a positioned leather thigh lacer or corset fixed to the uprights of a wooden crutch, the foot usually dangling below the corset. The child could ambulate with such a device or appliance without too much difficulty, but accurate comfortable fitting about the shortened extremity, particularly the weight bearing areas, was lacking and alignment was as straight as the crutch. The cosmetic value of such a rather crude device, without question, is poor. A child can get by and wear almost anything with the minimum of irritation and complaints. Such a child so fitted, however, lacks much to be desired: real comfort from a proper fitting corset, stability through improved alignment, comfortable support of all parts of the extremity including the foot, and functional value. A simple stick or a crutch tip does not allow such a youngster to wear a pair of shoes, does not provide stability for the foot and prevents such a youngster from achieving such physical feats as skating, etc. As the child prosthetic program developed at the Minneapolis Unit of The Shriner's Hospital for Crippled Children, it became obvious that the child amputee was more comfortable, had better function and looked better than a youngster with proximal femoral phocomelia using a crude pylon. The parents of the latter began to look at the lower extremity amputee child in the waiting room with envy. We had already begun with some success to utilize some of the improved prosthetic techniques in fitting, material and alignment for bracing paraplegics. The products looked better with closer fitting tolerances, prevented skin problems and improved function.

With this background, we began to develop "Pylon-Prosthetic" devices for the youngsters with sensible lower extremity congenital skeletal limb deficiencies, utilizing the already developed prosthetic techniques for support and bracing, alignment for improved stability and prosthetic ankle-foot components. Plastics, using total contact techniques, with and without metal reinforcement, have been used. The proximal weight bearing areas have been modified to surround redundant soft tissue as is present with proximal femoral phocomelia. Trap doors of various sizes and shapes have been necessary to allow placement of the device. Feet, as present, have been incorporated into the so-called socket, usually in as much equinus as is possible for improved cosmetic value. Alignment is achieved below the so-called supporting socket. In some cases, the remaining portion of the extremity is short enough to allow insertion of a knee joint mechanism and with such a device, so-called dynamic alignment is actually achieved.

Paraxial, fibular, hemimelias with relatively normal feet are quite adaptable to this technique. Usually the involved lower leg is shorter than normal and quite often the thigh is likewise, thus allowing application of a foot-ankle prosthetic component below the shortened extremity, with the natural foot-ankle in a position of relative equinus with good device alignment and the advantages to the child as described above.

Utilization of this technique, of course, is more expensive, but the advantages, we feel, support the added expense. Without question, the children exhibit happy satisfaction with their improved appearance, comfort, and increased functional ability. All of these children have voluntarily expressed their delight in being able to wear a pair of shoes, skate and perform other weight bearing tasks that they could not do before and in being like their normal friends. The parents display the same enthusiasm as the parents of the more standard child amputee.

Technical Aspects

Fitting children with congenital deformities is a challenging, and probably the most rewarding, area of prosthetics and orthotics.

One of the first things we must be aware of is that two similar cases may have completely different prosthetic solutions. Children with deformities of the type described may be fitted as early as six months, when the child starts to pull himself up to a standing position, or at any age thereafter. As the foot is in relatively satisfactory functional position amputation is not considered. Without amputation, harnessing is eliminated or simplified as the foot and leg "key" into the socket and provide satisfactory, stabilizing control of the "Pylon-Prosthesis." The parents of the child are grateful that no amputation is necessary and at night the youngster can ambulate on a functional foot for self-care functions with relative ease, without having to put on the appliance.

The clinic team prescribes each device after clinical and x-ray evaluation.

If the child is able to stand for cast fitting, primary pressure is applied to the foot of the affected side. If the child is able to cooperate in a standing position blocks are placed under the affected side, adjusted to the child's balanced standing posture. If knee locking strength will support weight, we may consider a below knee type "Pylon-Prosthesis." Major weight bearing will be on the plantar aspect of the foot in equinus position. The foot in equinus in line with the lower leg or tibia provides a very acceptable cosmetic result. Knee instability in several cases required knee bracing, joints, and lacer similar to a standard below knee fitting. Added bracing may later be removed as knee stability improves. In the event ischial support is necessary, the socket is extended superiorly or toward the pelvis and flared at the posterior-medial aspect for ischial support. Sockets also have been extended over the buttock for additional support.

Casts are taken with the knee in a neutral, relaxed position. This is important in the prevention of concentrated pressure areas within the socket which cause skin injuries.

Casts are modified by molding for displacement of soft tissue under load conditions. A plaster check socket is then made and fitted. Major corrections made at this time include relief for bony prominences, filling areas where there is no contact, and a trap door or window to facilitate application and removal of the test socket, which ultimately is incorporated in the final socket. There should be adequate space for the foot in the equinus position, particularly the toes, for the free motion required to prevent toe blistering or callous formation. There should be additional space anterior of or distal to the toes to provide some degree of space for growth, as experience indicates that crowding due to growth usually occurs within the foot portion of the socket long before it is significantly noted about the extremity above. The medial malleolus prominence usually requires extra spotting.

When the check socket appears to fit properly and can be easily applied and removed, the appliance below the socket is completed. If there is sufficient clearance between the foot portion of the socket and the floor, an adjustable leg with properly sized foot is installed for completion of the alignment process. An angle iron is used in lieu of the adjustable leg if there is insufficient space for adjustable leg techniques. Alignment with an angle iron is carried out in two planes with the child bearing weight to the floor via the angle iron which is moved about until the maximum balance point is found. Plumb lines are then used as reference points to complete this type of alignment procedure. When the adjustable leg alignment method is used the usual transfer techniques complete the appliance. The extension portion of the "Pylon-Prosthesis" is usually fabricated from lightweight wood, reinforced with a laminate.

These devices are serviceable from 12 to 24 months. The major factor determining appliance life is growth.

Case Presentation

The following is a summary of the cases at Minneapolis Unit, Shriner's Hospital for Crippled Children, who have been provided with so-called "Pylon-Prosthetic" devices with five illustrated cases.

Femoral Deficiencies

I. Femoral deficiency, distal femoral epiphysis, congenital malformation with knee dislocation, left. One case.

Case #1

Male infant, born in 1958 was admitted to Shriner's Hospital in 1959. The left knee was explored, a mal-formed semi-absent distal femoral epiphysis observed and the knee dislocation reduced. The patient was fitted with a crutch-brace type pylon in 1960. Derotation osteotomy to improve alignment was carried out in 1963. Fibrous ankylosis of the knee joint by that time had occurred. The youngster was then fitted with a so-called "Pylon-Prosthetic" device in 1963, replaced because of growth in 1964. The youngster is wearing a regular shoe and for practical purposes is as active as any youngster, age 7, could be, able to skate and engage in competitive sports consistent with his age. (Fig. 1-A through E).

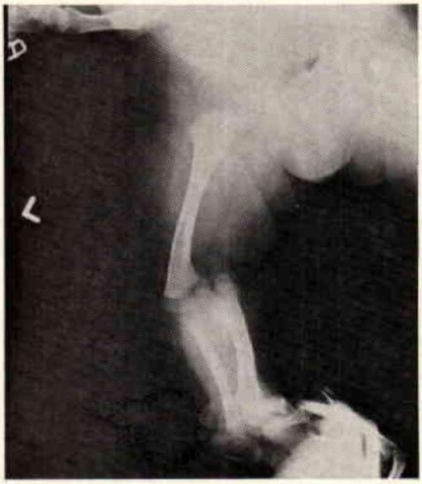

FIGURE 1-A: Case 1. Anterior-posterior roentgenogram showing knee deformity.

FIGURE 1-B: Case 1. Anterior photograph of deformity.

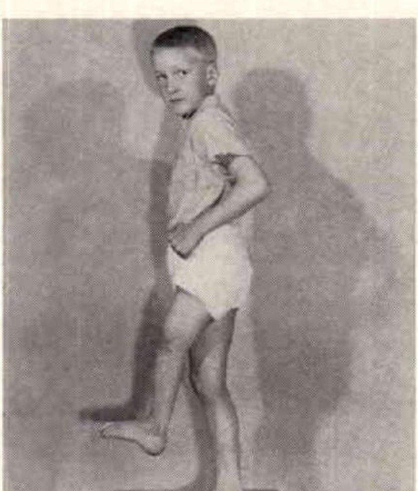

FIGURE 1-C: Case 1. Lateral photograph of deformity.

FIGURE 1-D: Case 1. Anterior photograph of "Pylon-Prosthesis."

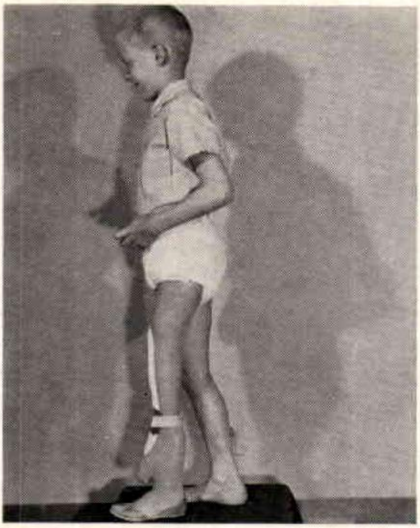

FIGURE 1-E: Case 1. Lateral photograph of "Pylon-Prosthesis."

FIGURE 2-A: Case 2. Anterior-posterior roentgenogram showing complete femoral phocomelia.

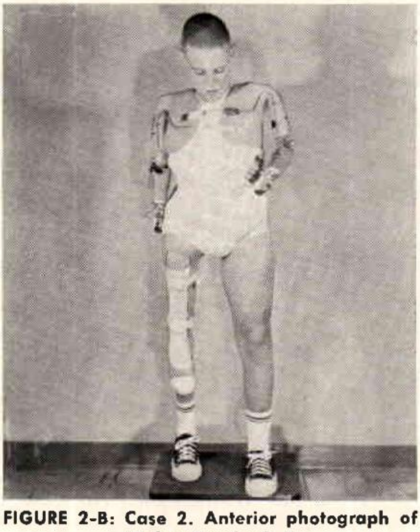

FIGURE 2-B: Case 2. Anterior photograph of "Pylon-Prosthesis."

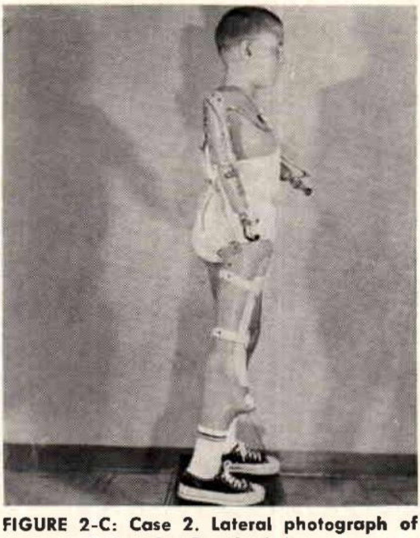

FIGURE 2-C: Case 2. Lateral photograph of "Pylon-Prosthesis."

II. Phocomelia, proximal, complete (femoral), right. One case.

Case #2

Male youngster, born in 1953 with phocomelia, femoral, proximal, complete, right. He was admitted to Shriner's Hospital in 1955 and fitted with a crutch type pylon. He was re-fitted with a plastic "Pylon-Prosthesis" in 1963, replaced in 1964 because of growth. This youngster also has bilateral upper extremity transverse phocomelia, fitted with bilateral shoulder disarticulation type prostheses with excellent upper extremity prosthetic function. His weight bearing balance and over-all functional ability has been much improved by the "Pylon-Prosthesis" in addition to cosmetic improvement. (Fig. 2-A through C).

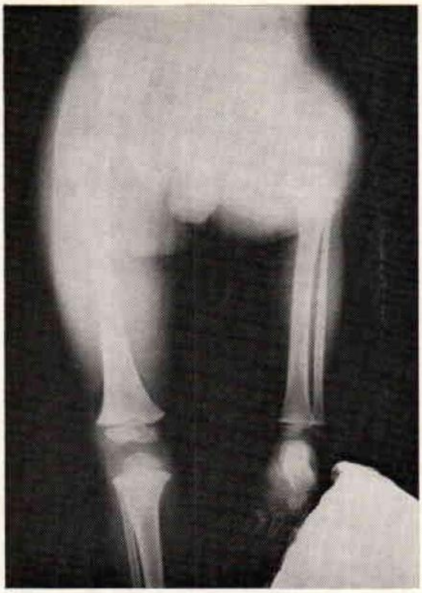

FIGURE 3-A: Case 3. Anterior-posterior roentgenogram showing incomplete femoral phocomelia.

FIGURE 3-B: Case 3. Photograph of shoe elevation.

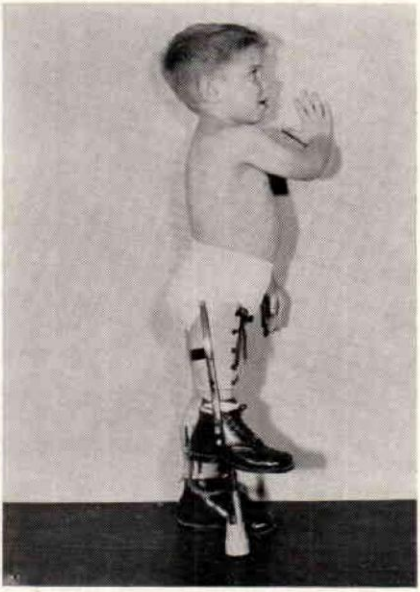

FIGURE 3-C: Case 3. Photograph of crutch tip brace.

FIGURE 3-D: Case 3. Anterior photograph of "Pylon-Prosthesis."

III. Phocomelia, proximal, incomplete (femoral). Two cases.

Case #3

Male youngster, born in 1955, with femoral phocomelia, proximal, incomplete, right, was admitted to Shriner's Hospital in 1958 wearing a high shoe elevation which was exchanged for a crutch type brace pylon. He was

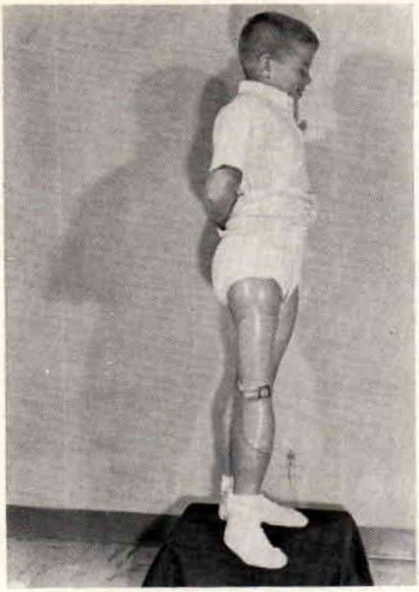

FIGURE 3-E: Case 3. Lateral photograph of "Pylon-Prosthesis."

fitted with a plastic "Pylon-Prosthesis" device in 1962, replaced in 1963 and again in 1965 because of growth. The plastic appliance has improved the patient's stability and allows him to wear a standard shoe and in turn has made it possible for him to engage in sports such as skating in addition to the cosmetic improvement. (Fig. 3-A through E).

IV. Proximal femoral focal deficiencies. Six cases (one bilateral).

Proximal femoral focal deficiency cases have been fitted with similar so-called "Pylon-Prosthetic" devices.

Four youngsters with femoral deficiencies have other significant skeletal deformities which include upper extremity transverse hemimelias and one with a lower extremity amelia. These other skeletal deformities have been replaced by functioning prosthetic devices.

Hemimelia, intercalary, paraxial, complete, fibula

Three cases of this deformity with relatively normal feet in relatively normal position have been fitted with plastic "Pylon-Prosthetic" devices with improvement of stability, comfort and cosmesis.

Case #4

This, the oldest case, age 8, wears a standard shoe and is able to participate in all play and sports consistent with his age. (Fig. 4-A through C).

Case #5

A male youngster, born in 1963, with right fibula complete, intercalary, paraxial hemimelia has been fitted with a rather unique plastic "Pylon-Prosthesis" device as illustrated. This device is extremely simple, light and easy to apply, very stable for the function necessary for a youngster of this age. (Fig. 5-A through E).

Summary

In summary, we have described plastic "Pylon-Prosthetic" devices for children with lower extremity congenital skeletal limb deficiencies, particularly applicable for cases of proximal femoral phocomelia and paraxial fibular hemimelia. These devices provide improved functional capabilities, comfort and cosmesis. The morale of the parents and the child with this affliction is much improved. These improvements, we feel, are well worth the added expense.

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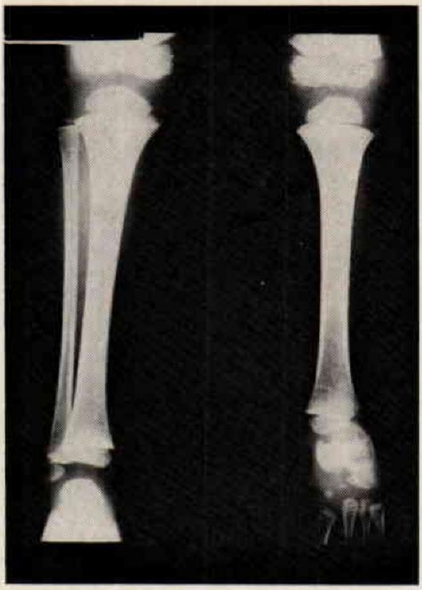

FIGURE 4-A: Case 4. Anterior-posterior roentgenogram of fibular hemimelia.

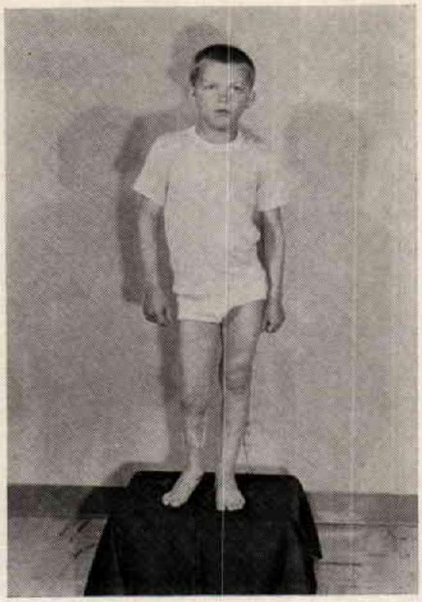

FIGURE 4-B: Case 4. Photograph of fibular hemimelia deformity.

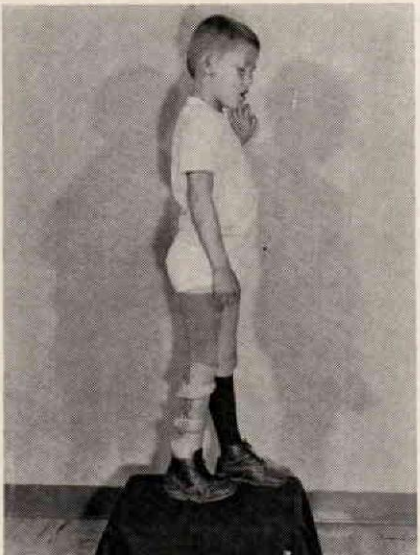

FIGURE 4-C: Case 4. Photograph of lower leg "Pylon-Prosthesis."

FIGURE 5-A: Case 5. Anterior-posterior roengenogram of fibular hemimelia.

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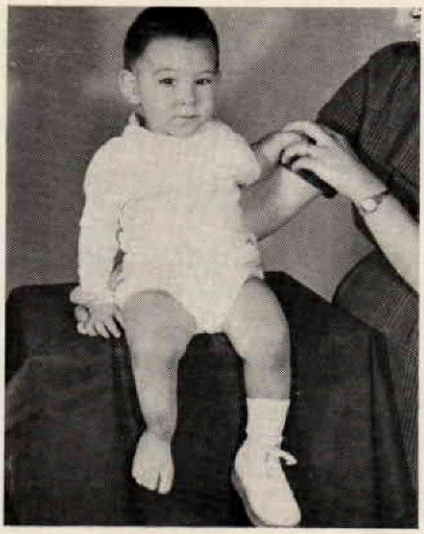

FIGURE 5-B: Case 5. Photograph of fibular hemimelia deformity.

FIGURE 5-C: Case 5. Photograph of lower leg "Pylon-Prosthesis."

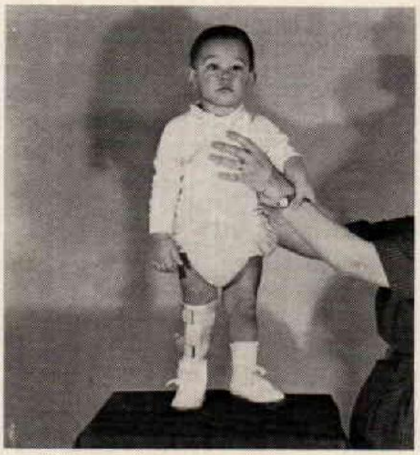

FIGURE 5-D: Case 5. Anterior photograph of lower leg "Pylon-Prosthesis."

FIGURE 5-E: Case 5. Posterior photograph of lower leg "Pylon-Prosthesis."

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