

THE KNUD JANSEN LECTURE

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The operative treatment of congenital limb malformation

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Introduction

First, I would like to express my thanks for the great honour paid to me in entrusting me with the Knud Jansen Lecture for the ISPO World Congress of 1980. Equally it is with great pleasure that I accept the proposed subject of the operative treatment of congenital limb malformations. I should say at the outset that the operation cannot be seen in isolation; it forms part of an over-all rehabilitative concept and is interwoven as part of the fabric of multi-disciplinary co-operation. In that sense, my contribution is at the same time an acknowledgement of the work of Knud Jansen who, for a number of years, was Chairman of the Committee on Prostheses, Braces and Technical Aids and later, acting as the first President of the ISPO, established multi-disciplinary cooperation as the basis for the activity of the ISPO. This same co-operation within the ISPO, has provided me with essential stimulus from many sources regarding both the philosophy and detailed practical procedures in the therapy of malformed children.

It follows therefore, that the complete clinic team is a prior desideratum for the successful comprehensive treatment of the child. In particular I wish to stress one main point, namely, the necessity of the mother or father's presence during the child's stay in hospital; this is especially important in the case of the baby and young infant. During this time the parent(s) is instructed and indeed, involved in both nursing procedures and therapy thus ensuring cooperation with the clinic team concerning the special needs of their child. We help the parents to find their way about the hospital and clinic and to understand the pre and post-operative measures thus obtaining their active participation.

Our experience over many years permits us to state with conviction that the child's postoperative need for analgesics and sedatives is considerably lower if the mother or father stays with the child in hospital. Since we insisted on the parents' presence, we have also observed a decrease in post-operative infections. While details cannot be given here, I remain convinced that the close co-operation with the parents is one of the main reasons for the happy, nay, joyful atmosphere in the paediatric service, for the mental health of the child in hospital and particularly for the long-term therapeutic success.

Operations are necessary if limb malformations demand functional or predominantly cosmetic corrections. In general, functional indications are widely accepted and disputed only in respect of timing and the specific operative technique. In the case of babies and young infants cosmetic considerations may assume paramount importance with the parents and especially so with the mother. However if concern for the appearance alone of, say, a club

hand determines the surgery then years later both parents and child will complain about the functional loss. As it happens functional loss may have a more profound and unfavourable influence on the development of the psyche than any departure from normality in appearance. Accordingly it is essential to spend sufficient time with the parents to understand their concerns and to ensure that they in turn acquire a real understanding of the proposed treatment and management.

New techniques allied to the sophisticated armamentarium of today have substantially improved the operative possibilities in hand surgery and the chances of success concerning malformations. So, before entering kindergarten or school, the child's stigma of malformation can be removed or at least be considerably alleviated from the outset.

Based on our knowledge and experience, we must carefully consider contraindication and indication, the precise type of surgery required and its timing, the selection of armamentarium as well as pre and post-operative treatment.

Now I want to explore some of these ideas with you and illustrate them by a few selected examples. A comprehensive description cannot be provided in the time available.

Acrosyndactylism

In acrosyndactylism, the constriction band syndrome, it is necessary to separate the peripheral and often overlapping syndactyly of shortened fingers at the earliest possible time. If left the constriction bands will delay and reduce growth and promote faulty development (Fig. 1 top left).

Initially it is not necessary to explore deeply because of the danger of relapse; this can be done later. At first, the fingers, even if they present in a stump-like form should be given the opportunity to grow in a straight way and to be used in grasping. We take advantage of the preformed commissures which are running in fine skin canaliculi (Fig. 1 top right). Thus, in acrosyndactylism we should in the first weeks of life separate all interconnections of the fingers of one hand, requiring only one operating session. If in addition there are deep constriction bands with peripheral oedema or even arterial blood supply disturbances, excision of the constricting connective tissue structures and Z-plasties should have priority in the operative planning (Fig. 1 bottom left and right).

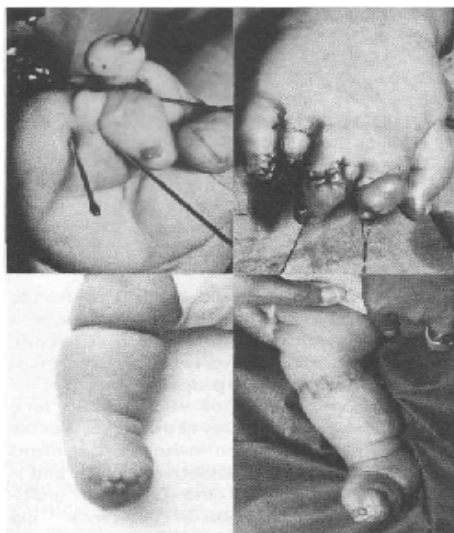


Fig. 1 Top left, typical acrosyndactylism of the right hand in a baby with a quadrilateral constriction band syndrome. The index, middle, ring and little fingers are short and grown together. At the start of the operation the small skin canaliculi are marked with a sterile blue dye. Top right, the same hand during the operation to separate the fingers. Bottom left, the same patient's right leg before surgery. Bottom right, about six weeks after excision of the constricting tissue structures with Z-plasty, oedema of the foot shows some reduction.

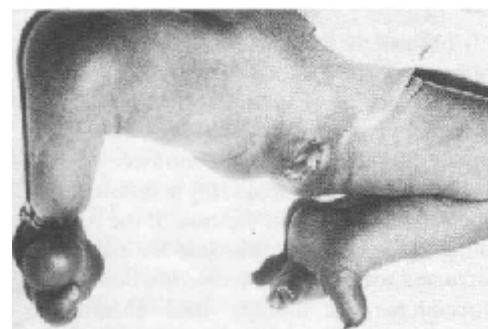


Fig. 2 Constriction band syndrome combined with atypical cleft feet. The oedema of the right foot is caused by the constriction band just above the ankle.

In connection with the constriction band syndrome, I wish to report on one particular child (Tamara, M.) who-apart from the fingers' acrosyndactylism presented with atypical cleft feet, constriction bands of the right leg and foot with peripheral oedema, and a congenital skin deficiency of the lateral right leg (Fig. 2). I first performed excision of the constriction band with a Z-plasty of the right lower limb, then, considering the situation, carried out an atypical disarticulation at the ankle joint to produce an end-bearing Syme-like stump end. As illustrated the cross hatched skin areas and the whole foot skeleton were excised (Fig. 3 top left). The muscle bellies were carefully prepared, cut and connected to the stump end; the skin was tailored and sutured over the stump end (Fig. 3 top right and bottom left). On the left side an atypical cleft foot with a dorsiflexion contracture of the first ray was present thus preventing the fitting of a shoe. The cleft was eliminated and the first metatarsal bone fused to its neighbouring ray. In this way the tibialis anterior muscle could now act on the whole foot. By these operative measures including the disconnection of the syndactylism, and in co-operation with prosthetic technique, the way was paved for a normal life at kindergarten and later at a normal school as well as providing for successful social integration (Fig. 3 bottom right).

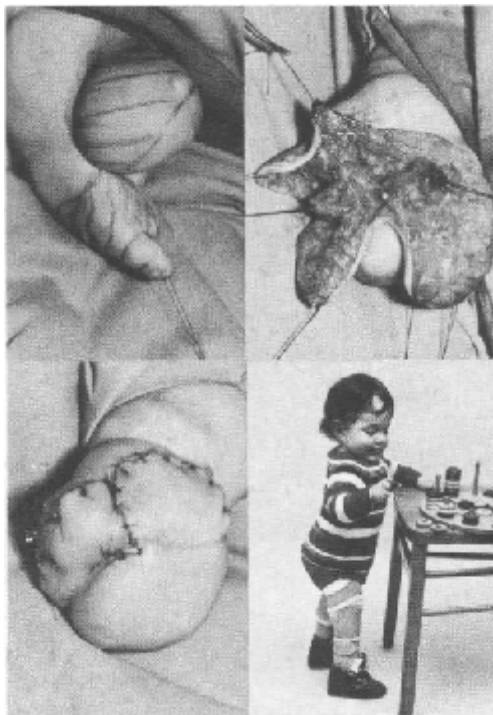


Fig. 3 Top left, right foot of patient shown in Figure 2, the cross hatched areas were excised. Top right and bottom left, atypical disarticulation of the ankle joint to produce an end bearing Syme-like stump (see text). Bottom right, the patient fitted with a Syme prosthesis on the right side and an orthopaedic "Innenschuh" on the left.

Endogenous syndactylism

Endogenous syndactylism does not require early operation before the child is weaned; but separation of the fingers should be completed before school entrance. Early operation, however, is required: 1) in the Apert's syndrome in order to set free the thumb; 2) if,

In the case of syndactylisms of the lower limbs, in general there is no indication for operative intervention, except where the skin canaliculi lead to hygienic problems.

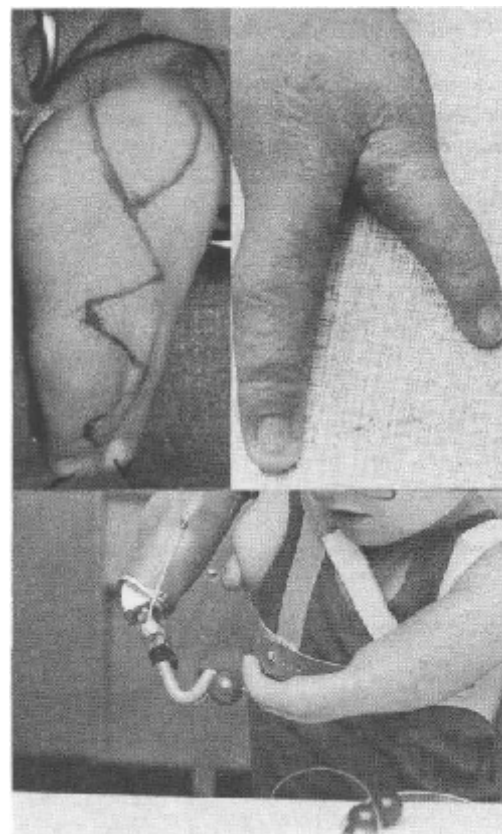


Fig. 4 Top left, the only two fingers of a baby with left phocomelia and right amelia. Top right, the same hand six years after surgery. Bottom, the patient demonstrating bimanual play. The right side is fitted with a short shoulder disarticulation prosthesis; note the functional "club hand" position of the hook

by differential growth of the mutually connected fingers, increasing contractures occur; 3) the condition only affects two fingers (Fig. 4 top left). This child presents with an amelia of the upper limb on the opposite side. In this case, I performed in the suckling period separation of the syndactylism in the little phocomelic hand following the technique of W. Blauth, permitting full use of the grasp between the fingers. Now six years after the separation of the syndactylism no tendency to recurrence can be seen (Fig. 4 top right). The child has achieved a very dexterous synergy of the two fingers, acting in concert with a short prosthesis with a hook on the other side (Fig. 4 bottom). The hook is disposed in the functional "club hand" position.

Longitudinal deficiencies, metacarpals and phalanges central

Cleft hands usually demonstrate excellent function and indeed are more difficult to recognize in motion than in a resting position (Fig. 5). There may be an indication for operation if the thumb is conjoined to the "index" in the manner of syndactylism. The emancipation of the thumb thus provided significantly improves function. In general there is no indication for operative intervention in the cleft hand or cleft foot (Tamara, M. (Fig. 2) is an exception).

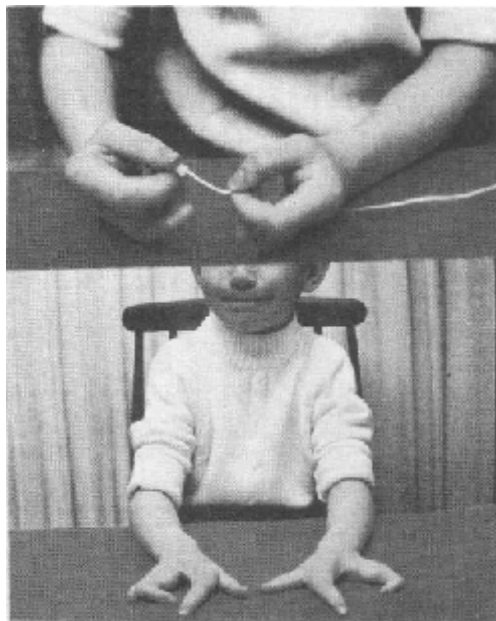


Fig. 5 Top, cleft hands during functional movement. Bottom, the same hands at rest

Polydactyilia and Macrodactyilia

Polydactyilia, duplication of the thumb, and macrodactyilia both require early operative treatment to avoid psychological damage from the abnormality. Operation is performed soon after birth and is now made easier with micro-surgical techniques. In the case of terminal double thumb (Fig. 6 top) which is a very frequent form of polydactyilia, the part with the smaller nail is excised. In our experience so far this has always been the radial part. When the nails are of equal size from the beginning the radial moiety is removed with the resultant scar on the outer side. If the medial one is removed then the

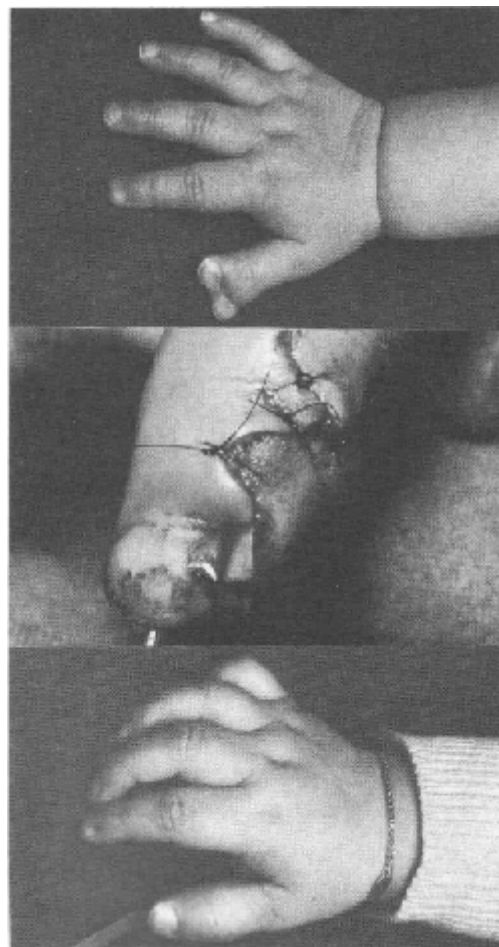


Fig. 6 Top, duplication of the distal phalanx of the thumb. Centre, during surgery the radial part is excised and the thumb stabilized by a longitudinal Kirschner wire. Centre, adaptive sutures are shown in situ before final preparation of the skin flaps and fine skin suturing. Bottom, the result six weeks after surgery.

scar might interfere with the important grasp between thumb and index. Important in this procedure are the diminution of the joint area at the interphalangeal joint, the need to construct a "ligament" on the radial side and the use of Kirschner wire for temporary "stabilization" (Fig. 6 centre). It is sometimes necessary to perform a corrective osteotomy of the proximal phalanx. After treatment includes the use of a temporary finger splint and, of course, physiotherapy (Fig. 6 bottom).

In case of macrodactylia, W. Blauth and G. Loesch recommend the resection of the hypertrophic nerves soon after birth. We have so far destroyed the relevant epiphyses and reduced the hypertrophic tissues as soon as the size of a corresponding finger of the father or mother had been reached. According to W. Blauth, despite many operations amputation is ultimately necessary except in the case of the thumb. In our view the loss of a single digit (except the thumb) can be well compensated. There is no doubt that a child with gigantism of a finger has to bear a lot of teasing and ridicule at both kindergarten and school and will experience the grief and frustration associated with the stigma of a physical disability even more intensely than an amputee.

Longitudinal deficiencies

In case of deficiency of the first metacarpal and the thumb but without club-hand, pollicisation of the forefinger is indicated. Dysplastic thumbs should not be pollicised; despite meticulous surgery failure can be expected. In such a case, it is better to remove the dysplastic thumb and most of the first metacarpal leaving the metacarpal base and pollicising the index finger eventually transposing it to the base of the first metacarpal. Until now I have delayed pollicisation until the age of four in order to ensure the necessary co-operation from the patient post-operatively. In contrast, Buck-Gramcko advises operation before the child is weaned. The operation is carried out according to current practice. The decision to proceed to operation must be based on a comprehensive test of the function of the whole hand. If the patient requires to use a walking stick to walk then pollicisation may be required urgently as in the case of a thalidomide damaged child in one case with additional tibia deficiency on both sides, a right sided femur varum and left sided proximal focal femoral deficiency. The pollicisation was performed according to the technique of Blauth (Fig. 7) which avoids the need for free skin grafts.

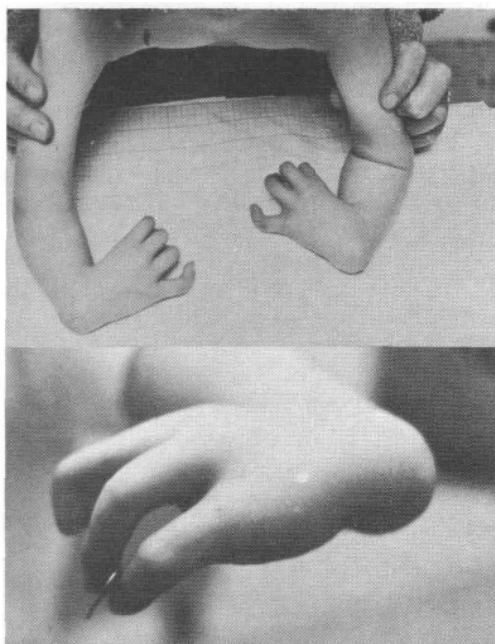


Fig. 8 Top, radial club hand due to bilateral total deficiency of the radius and ray I. Bottom, the little finger compensates for the thumb

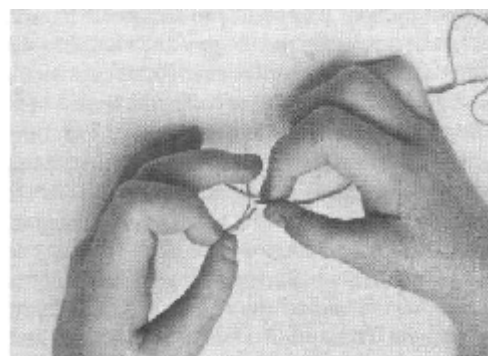


Fig. 7 Result of bilateral pollicisation of the index fingers using Blauth's technique

The radial club-hand results from a longitudinal deficiency of the radius, partial or total, and is usually accompanied by a right-angled radial abduction contracture, a palmar flexion contracture, a dysplasia or even complete asplasia of the first metacarpal and thumb and with contractures of the other fingers decreasing from the radial to the ulnar side. Additionally, the concerned limb often shows significant contractures at both elbow and shoulder joints (Fig. 8 top). The arm is thus restricted in its entire function. As a result the radial club hand is directed towards the body and is able to accommodate very well to many of the activities of daily living. In the club hand, the little finger compensates for the thumb (Fig. 8 bottom) closing of the "fist" is achieved by extreme radial abduction and palmar flexion so that the hand touches the forearm. The absent pronation and supination is compensated for by movements at the abnormal wrist joint and the flabelliform nature of the metacarpals (Fig. 9 top left and right); the reduced flexion at the elbow and internal rotation at the shoulder are functionally balanced by the radial abduction of the hand (Fig. 9 bottom).

Despite these functional compensations the first sight of the club hand causes a serious psychic shock in the mother. So it is understandable that the parents, in their distress, try to find a surgeon who can be persuaded to remove this ugly and very disturbing malformation. Early operations, however, often lead to severe losses of function, as a result of growth and ankylosis of the hand joint in a functionally unfavourable position, and, if both limbs are concerned, to helplessness and permanent dependency on others (Fig. 10). It is essential that immediately after birth physiotherapy is started with the objective of overcoming or at least, reducing contractures and of increasing the range of movement and thereby the overall function of the limb.

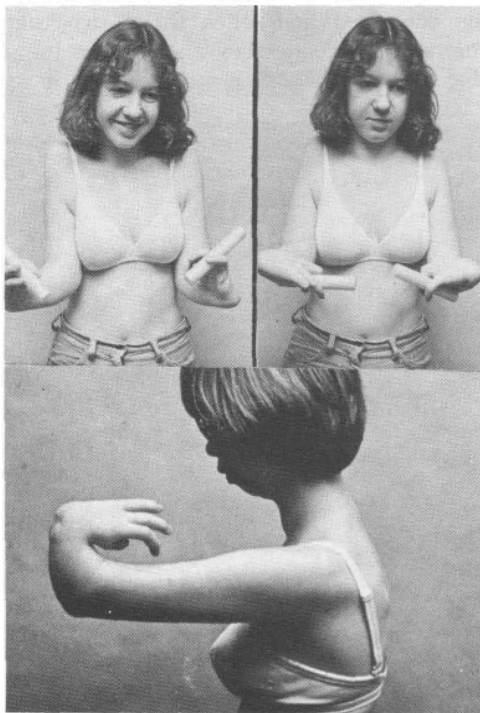


Fig. 9 Radial club hand; top, compensatory movements for pronation and supination of the forearm and for internal rotation of the shoulder joints. Bottom, the radial abduction compensates for loss of elbow function.

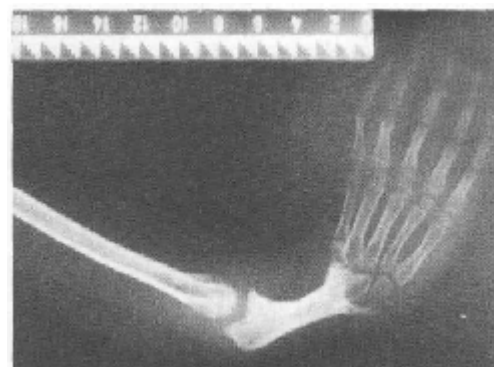


Fig. 10 Arthrodesis of the wrist and destroyed growth following corrective surgery of the club hand in early childhood.

The primary treatment starts directly after birth and consists of passive stretching which is repeated as required by the mother four to five times each day when diapering the child with supervision of the therapist from time to time (Fig. 11 top). The club hand is thereby gradually brought into the normal position with the distal end of the ulna serving as the fulcrum (Fig. 11 centre). The position achieved is at first held in a plaster of Paris splint and from the third or fourth month of life by a PVC splint with an adjustable leather strap (Fig. 11 bottom). The splint should not impede the mobility of the fingers and from the fourth month of life is only worn during the night. Passive stretching is gradually replaced by active exercises. Thus, in many cases we may succeed in bringing about a complete passive correction of the club hand and in considerably increasing the hand's range of movements, but we can almost never succeed in actively and totally maintaining the position achieved. The total correction by graduated passive stretching represents the ideal preparation for corrective surgery the indication for which must be strictly observed. Our practice is that hand and arm function must be carefully tested and these tests must prove that no functional losses in either hand or arm or in bimanual tasks will accrue from the operation. No such functional loss should occur if a unilateral club hand is such that the normal or less seriously damaged hand of the opposite side can compensate for any functional deficit resulting from operation.

Embracing these strictures I do not operate before the end of the growth period and use a technique which preserves the mobility between the two rows of carpal bones. In the case of a total radial deficiency the ulna is resected distally to an amount determined by radiographic study. The object is to fuse the distal ulna to the lunate and triquetrium suitably denuded of articular cartilage on their proximal aspects. In this way a broad support is provided which will serve to prevent recurrence of deformity without disturbing the intercarpal "joint" in its important function; furthermore, by this procedure the important fan-like movements of the metacarpals are preserved. Thus it is worthwhile continuing the conservative treatment in close co-operation, at first with the parents and later increasingly with the disabled themselves until the end of growth, whether the patient decides for the operation or rejects it.

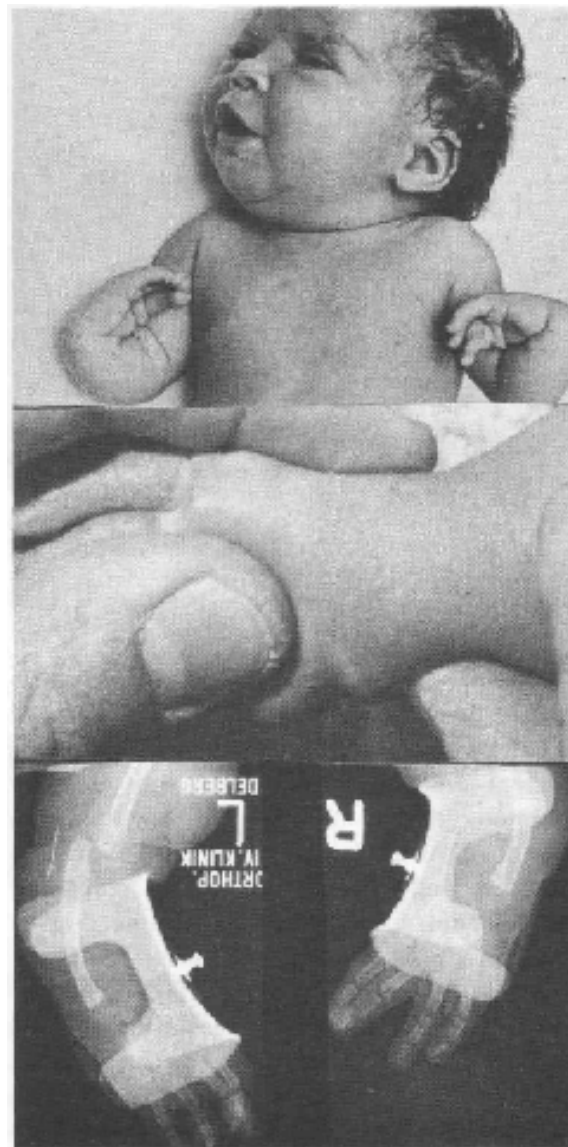


Fig. 11 Top, new born child with club hands due to bilateral total longitudinal deficiency of the radius and ray I. Centre, passive stretching, using the distal ulna as a fulcrum, should be started on the first day after birth. Bottom, club hands with plastic splints in situ six weeks after the start of treatment (see text). (Figure 11 top and bottom are from *Rehabilitation Monograph 44, The total treatment of the limb deficient child, Institute of Rehabilitation Medicine, New York University.*)

Once more I would like to underline the great importance of the flabellate movements of the metacarpals in compensating for a radio-ulnar synostosis. It is for this reason I operate on radio-ulnar synostoses only if the attitude of the hand is in a functionally unfavourable over-pronation; then I perform a rotation osteotomy in the proximal radio-ulnar union and promote the flabellate movements of the metacarpals after consolidation.

The *Longitudinal deficiency of ulna*, total or partial is with regard to function more difficult to judge than the radial one, especially if it is combined with a humero-radial synostosis. As a

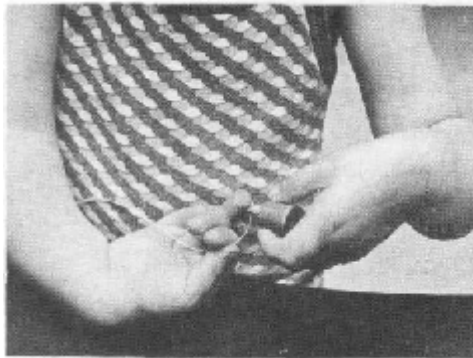


Fig. 12 Loss of function in total or partial longitudinal deficiency of the ulna is more severe than in the radial deficiency; the dorsum of the hand is lying on the table because of the fixed supination of the forearm.

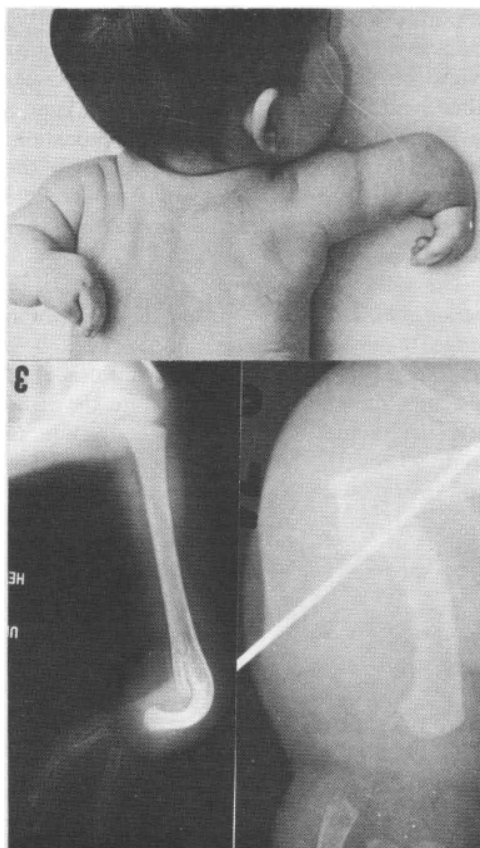


Fig. 13 Top, total longitudinal deficiency of the ulna, ray IV and ray V with bilateral humero-radial synostosis. Both shoulder joints are internally rotated and the congenitally fused elbows are hyperextended. Bottom left, radiograph of left arm. Bottom right, surgical transposition of the left forearm from hyperextension to flexion following osteotomy

result of the supination contracture the hand presents to the middle of the body, but, in contrast to the radial club hand, with an upwardly directed palm, the dorsum of the hand lying on the table is an unfavourable position for most tasks (Fig. 12). Moreover, in case of the humero-radial synostosis both forearm and hand often face backwards because of hyperextension of the elbow and/or an internal rotation contracture of the shoulder (Fig. 13 top and bottom left). If both sides are involved it is recommended to provide function by performing a rotation osteotomy of the humerus on one side and the considerably more difficult flexion osteotomy in the area of synostosis on the other (Fig. 13 bottom right). Afterwards, the hand of the rotated side is in supination and the other, after flexion osteotomy, in pronation. Only by these measures can the hand-in-hand activities of work, play and bodily functions be performed. Thus the pronated hand is on the table for writing and working while for example, the supinated hand can be used for cleaning after defecation (Fig. 14). However, the basic approach remains a conservative one, in accordance with the views expressed in the management of the radial club hand. Operations that correct the ulnar club hand are generally not necessary.

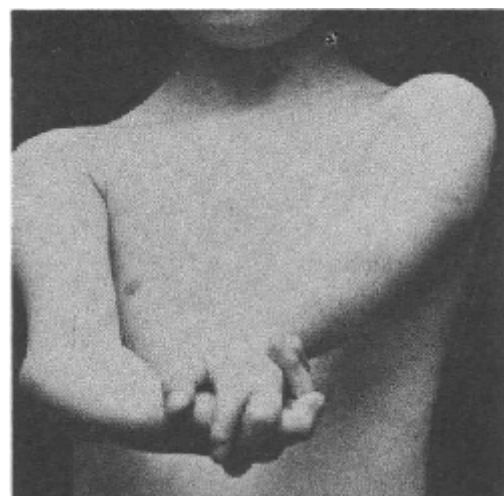


Fig. 14 Same patient shown in Figure 13 five years after operation. The left hand is positioned in pronation but the fused elbow is now more extended. The right, non-operated side, is still in supination; hand-to-hand activity is still possible.

Transverse deficiencies

The *congenital metacarpal, total or partial* is efficient and can be fitted well by a grasp abutment device or dolly or an open-end prosthesis. Additionally, a cosmetic glove can be provided at the appropriate point of development. Contractures which spread apart the first and fifth metacarpals as found in the severe form of symbrachydactylia require corrective osteotomy of the first and fifth rays together with excision of the central bones of the hand to construct, as it were, a cleft hand. In this way function is improved and it becomes possible to fit a cosmetic glove. In bilateral cases, a sensitive grasp and holding function should be established by surgery on one side at least (Fig. 15 left).

The *congenital amputation, forearm, lower 1/3, middle 1/3, upper 1/3* does not normally require surgical intervention. In bilateral cases of long to medium stumps, however, the

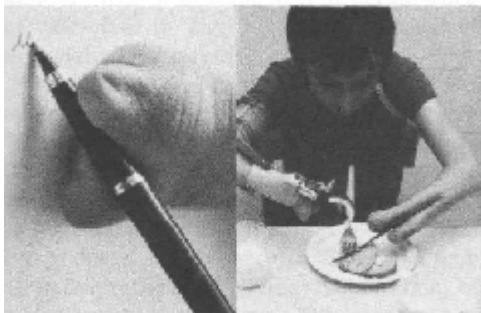


Fig. 15 Left, surgical deepening of the cleft between the first and fifth rays enables the patient to hold a pen or pencil. Right, a 15 year old patient with bilateral total transverse carpal deficiency. The right side is fitted with an open end prosthesis; the Krukenberg operation performed on the left side permits the patient to hold the knife, by means of a special adaptive device, with normal tactile sensation and force control.

forming of a "forceps" grasp with good sensation by separating the radius and ulna (Krukenberg, 1917) is particularly helpful, especially for the activities of daily living (Fig. 15 right). In my opinion there is one absolute indication for the Krukenberg procedure and that is in the case of the traumatic blind bilateral forearm amputee. Normally the Krukenberg operation is performed on only one side. After adequate physiotherapy and ergotherapy an ideal synergy between the sensitive Krukenberg grasping arm and an active forearm prosthesis fitted to the other stump can be achieved. Regarding the operation technical details I would refer you to the "Atlas of Limb Prosthetics: Surgical and Prosthetic Principles" soon to be published by C. V. Mosby.

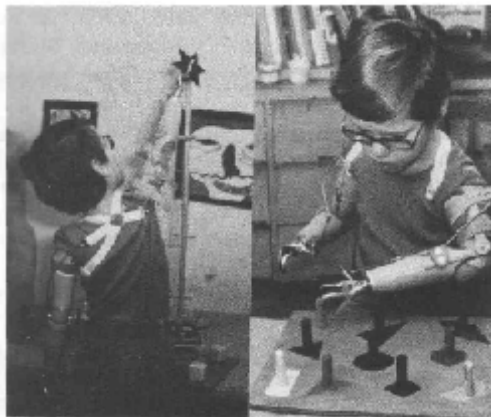


Fig. 16 A ten year old boy with bilateral transverse deficiencies comprising deficiency of the middle third of the right arm and total forearm deficiency on the left. Following bilateral angulation osteotomies of the distal humeral stumps he uses bodypowered prostheses with special fittings. Left, shoulder movements are transferred direct to the prosthesis. Right, the skin of the longer left stump is free for touching if the Hosmer locking elbow is flexed

The *congenital amputation, forearm, total* can be fitted well without operation and the prosthesis is self suspending as one can obtain an intimate fit at the epicondyles of the humerus by careful modelling. In case of hypoplastic epicondyles and in the case of arm lower 1/3 to middle 1/3 angulation osteotomy of the distal humeral stump (E. Marquardt, 1972) can be employed in order to achieve excellent distal suspension and rotational stability of the prosthesis with increasing effectiveness in prosthetic use (Fig. 16). After this operation the total range of motion at the shoulder joint and shoulder girdle can be utilized and this is particularly important in the bilateral case. Details of the operative procedure can be gleaned from the Atlas referred to. Another important indication for the angulation osteotomy is threatened perforation of the bone end through the skin due to osseous overgrowth in the lower to middle third of the arm

(Fig. 17). When there is danger of osseous overgrowth in the middle 1/3 to upper 1/3 of the

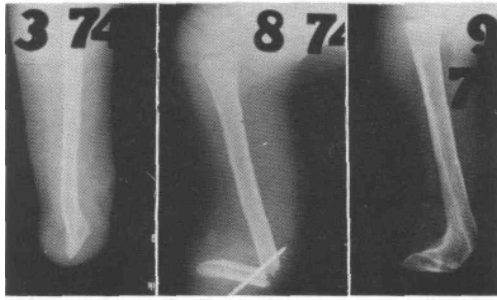


Fig. 17 Transformation of the bone structure following angulation osteotomy due to osseous overgrowth. After consolidation the Kirschner wire is removed; daily endbearing training and the use of an active prosthesis are necessary. (This figure is from *Z. Orthop.* 117 (1979) 622-331, F. Enke-Verlag, Stuttgart.)

arm "capping" of the bone end can be performed. Swanson and Meyer use a silastic end-cap and I use a chondro-osseous graft. Should the spike of bone have already perforated the skin there is no alternative but to shorten the humerus. Stump "capping" should be performed if perforation threatens again.

Case Study

The patient was born on 13 May 1965 with bilateral transverse upper arm 1/3 deficiency. Malformations of the lower limbs consisted of bilateral longitudinal femur subtotal deficiencies, bilateral fibula total deficiencies and bilateral ray (metatarsal and phalangeal) IV and V deficiencies. She suffered recurrent incidents of osseous overgrowth of the humerus on both sides and between 1969 and 1973 six reamputations were performed for this reason. Stump capping procedures employing autogenous cartilage-bone transplants were successfully carried out on each humeral stump in 1974.

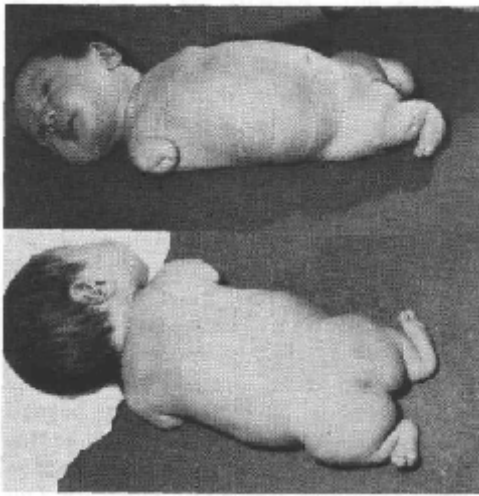


Fig. 18 The new born child showing the congenital scar at the end of the short above-elbow stump and extreme pes valgus. Left and right sides are almost identical.

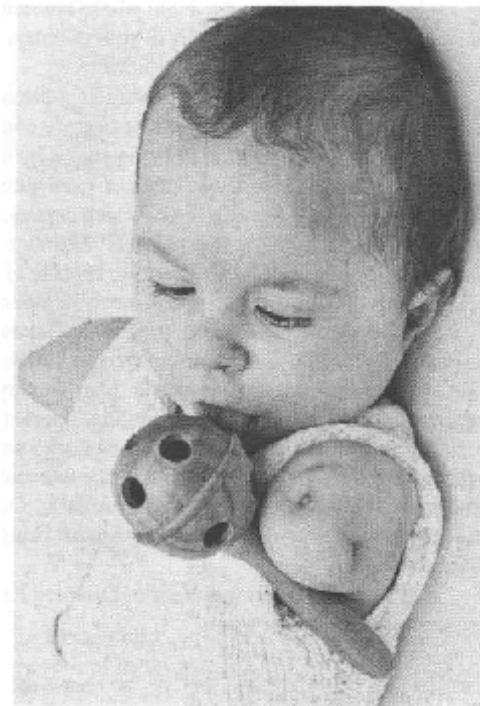


Fig. 19 The lower limbs were placed in a plaster of Paris cast; an orthosis was fitted at the age of three months. During this time the child used the stumps, chin and mouth for touching and grasping.

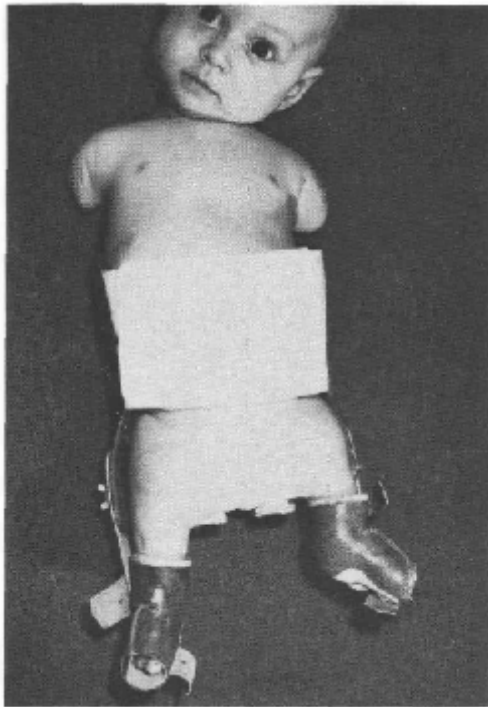


Fig. 20 The orthosis showing the orthopaedic leather shoes and metal foot supports. The orthosis could be extended while maintaining the feet in the optimum corrective position.

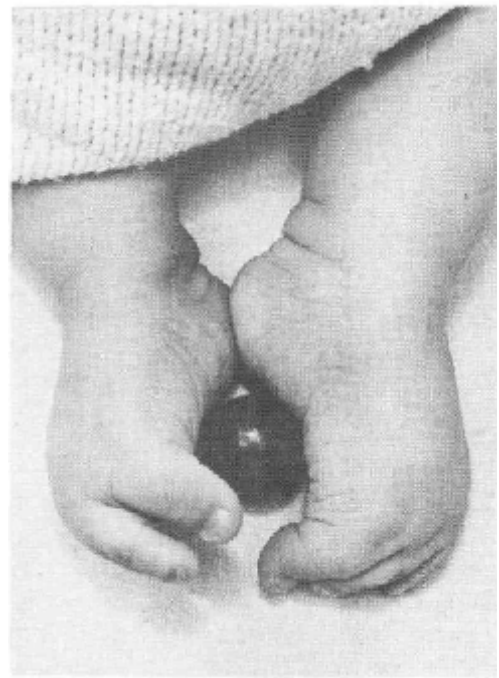


Fig. 21 The active use of both feet for play and motivated exercise is very important for the correction of deformity and the development of foot function.

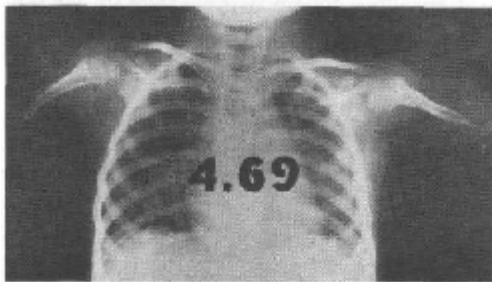


Fig. 22 The X-ray of the upper limbs shows osseous overgrowth in this congenital case which is identical to that seen in children following traumatic amputation.

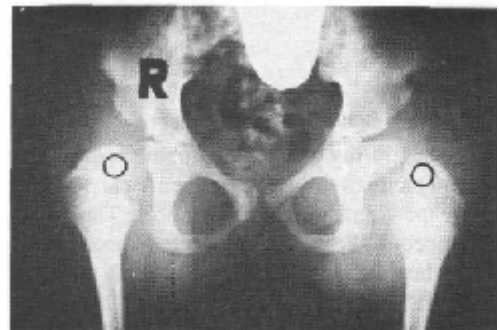


Fig. 23 The X-ray of the pelvis shows a rudiment of the femoral head in each hip joint and femoral elements in the form of a cap in synostosis with the proximal tibia bilateral. The fibula deficiency bilateral, is complete. The circled areas indicate the sites used for the cartilage-bone transplants



Fig. 24 Between 1969 and 1973 the patient endured six reamputations because of osseous overgrowth. The illustration shows a simple method of skin traction to

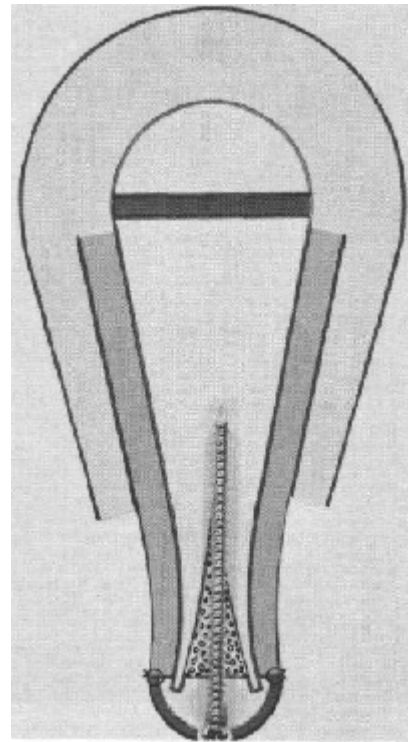


Fig. 25 Sketch of the stump capping procedure using a cartilage covered cap. The distal end of the humerus is split into two pillars which are

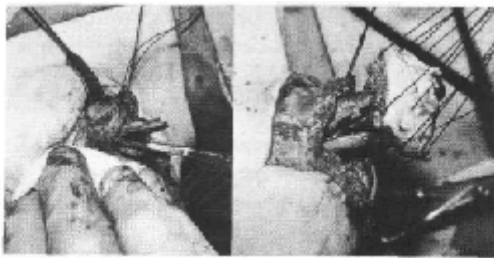


Fig. 26 Stages during the stump capping operation. The pictures show the procedure carried out on the right side in March 1974. The same procedure had been performed successfully on the left side in January 1974. Left, the end of the humerus is split and the musculoperiosteal flaps prepared for fixation to the transplant. Right, the transplant is fixed by a screw. In this case the pillars proved to be too long and the transplant was removed. The pillars were shortened by about 1 cm, the transplant was replaced and the musculoperiosteal flaps could then be attached to the transplant.

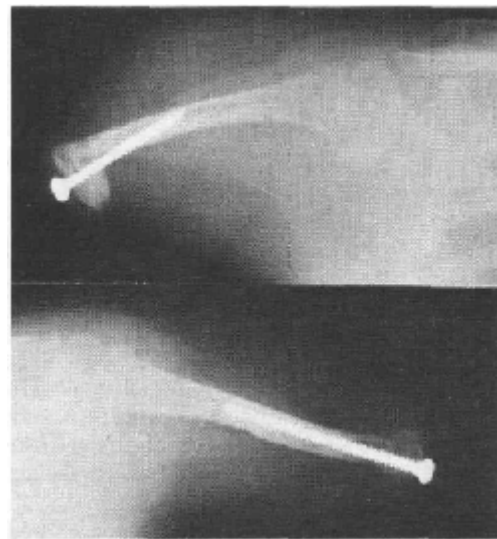


Fig. 27 Right humerus (top) and left humerus (bottom) in December 1974 just prior to removal of the screws. The transplant on the right side has been fashioned to provide better prosthetic fixation. Thickness of the cartilage is indicated by the distance between the distal end of the screw and the bone.



Fig. 28 Daily end-bearing training is essential after the stump capping procedure to stimulate normal development of the humerus and to prevent osseous

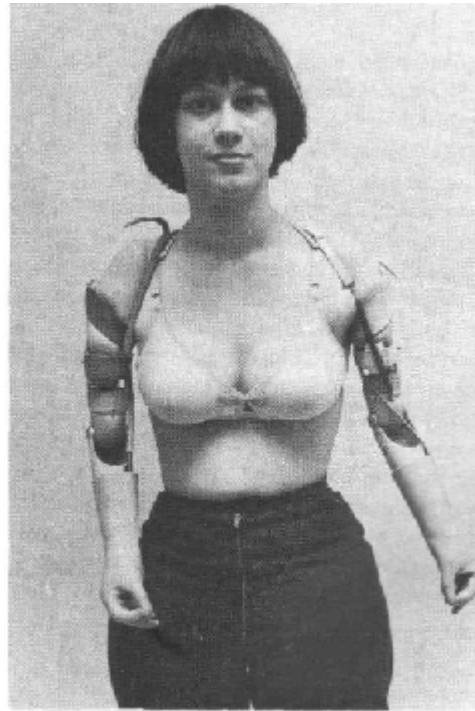


Fig. 29 It is also essential that the patient be encouraged to make daily use of active prostheses. Those shown are body powered with open socket construction to accommodate the bulky stump end. The length of the arms is a compromise; the patient's height varies as she does not always wear her extension prostheses.

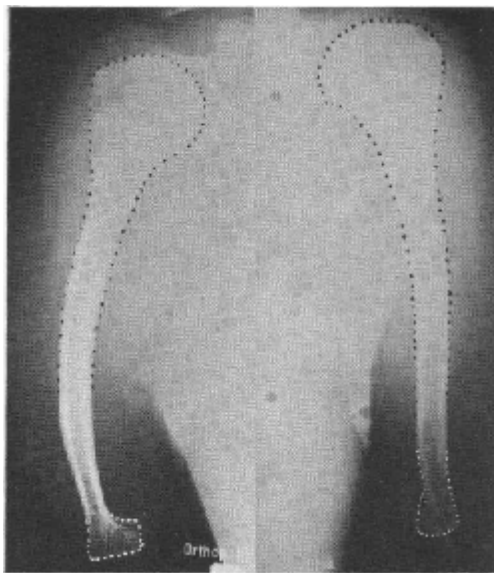


Fig. 30 The patient was last X-rayed in 1976; the pictures demonstrate good growth of both humeral stumps amounting to 3-4 cm on the right side and 3-2 cm on the left since December 1974.



Fig. 31 The capping procedure has prevented a recurrence of osseous overgrowth and produced almost normal development. Note the pterygium and the absence of axillary hair on the right side.

In the case of a patient who had not suffered perforation of the bone through the skin and reamputation, the incision for the stump capping procedure would be proximal to the stump end to provide a scar-free end bearing area. However up to November 1980 the patient has experienced no problems with her stumps.

In certain malformations of the lower limbs it is necessary to intervene surgically to make it possible for the child to walk, or to improve its walking ability. Surgical intervention is carried out at an appropriate stage in the child's development and in close co-operation with the prosthetist and orthotist.

Longitudinal deficiency tibia, total or partial

Orthopaedic-surgical treatment cannot be avoided if a child with this condition is to walk. If possible the operation is carried out at the suckling age, so that walking can start according to the child's development.

If, in the case of longitudinal deficiency tibia total, the femoral condyles and the knee capsule are normal, and if the child is to be operated on not later than in his second year of life, the Brown (1965) procedure—that is the construction of a knee joint between the femoral condyles and the head of the fibula—is indicated.

During a second operating session the distal end of the fibula is fused with the astragalus or calcaneus according to Blauth (1978) in preference to disarticulation of the ankle joint. From the third year of life, disarticulation of the knee joint is the method of choice if the tibia is totally lacking and a normal femur is present.

If the distal femur is hypoplastic, there may be a more or less serious disturbance of the growth and, after a knee disarticulation, a cone shaped, eventually mid-thigh and poor load carrying stump end. In such a case, especially if malformations of the upper limbs are

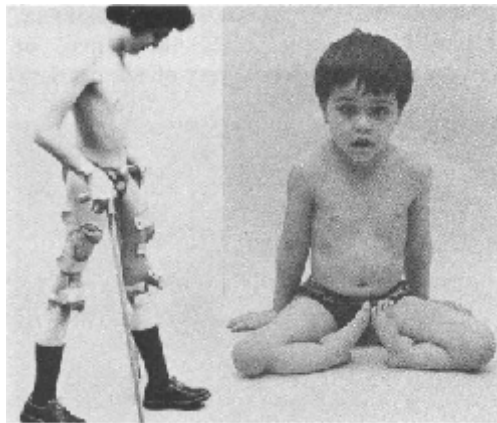


Fig. 32 Left, patient with longitudinal deficiency tibia, total bilateral, right coxa vara, PFFD type Aitken A left with sub-trochanteric pseudarthrosis; longitudinal deficiencies of metacarpal and phalanges total. The left hip was reconstructed and both knees and ankles fused. Patient is ambulatory (for case report see "Atlas of limb prosthetics", 634-637). Right, patient V.F. was non-ambulatory up to his 4th year because of bilateral tibial deficiencies.

present, fusion between the condyles of the femur and the head of the fibula is recommended; also between the distal end of the fibula and the astragalus or calcaneus, if necessary with ensuing partial amputation of the foot (Marquardt, 1981). Advantages in comparison with knee disarticulation are the better end bearing capacity and the self-supporting and rotation-stable fixation of the orthoprosthesis with Velcro closures which a child with a hand or arm disability may manage more easily than a knee disarticulation prosthesis (Fig. 32, left).

In cases of longitudinal deficiency of the tibia partial there is a better chance of success if the patient is operated upon as a baby, but even for the young infant there is a good chance that a load carrying leg with a functioning knee joint can be provided by surgery. Correct interpretation of the radiological signs is essential as the proximal epiphys is of the tibia is often not visible on the

X-ray (Jones et al, 1978). Meyer (1980) osteotomizes the fibula subperiosteally, preserves the periosteal tube and merges the distal fibular fragment with the chondric tibial rudiment. In Heidelberg, I transfer the proximal epiphysis of the fibula to a central position below the tibia rudimentum, stabilize with Kirschner wires and join the shaped proximal epiphysis of the fibula to a manchette of the periosteum of the tibia rudimentum and to the patellar ligament (Fig. 32, right and Figs. 33-37). For larger tibial rudiments which can be seen on the X-ray we osteotomize the fibula closely proximal to the level of the distal end of the tibia and fuse the distal fibular fragment to the tibial stump (Fig. 38).

Longitudinal deficiency fibula, total or partial

Accompanying malformations are shortening, bowing of the tibia, pes equino-valgus, fibular ray deficiencies of the foot and synostosis between the astragalus and calcaneus. Moreover, in some cases there may be disturbance of the lateral part of the proximal growing plate of the tibia. There is often a dimple-like skin retraction above the bowing of the tibia, a shortening of the triceps surae, often with fibrous tissue similar to the musculus sternocleidomastoideus in torticollis, and connective tissue tending to contracture as a result of being a rudimentum of a non-ossified fibular-anlage.

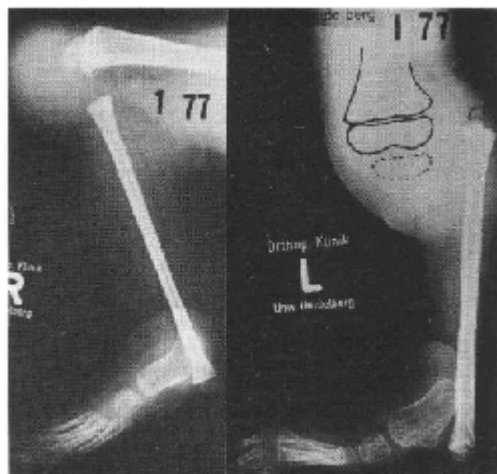


Fig. 33 Left, patient V.F. The X-ray of the right leg shows a longitudinal deficiency tibia, total. Type Ia (Jones et al, 1978). The Brown procedure is contraindicated because of the patient's age and the hypoplastic distal femur. Indication is for knee and ankle fusion or, as done, supracondylar osteotomy for correction of the severe flexion contracture and for fitting with orthoprosthesis. Right, X-ray of the left leg shows a longitudinal deficiency tibia total, but with almost normal femoral condyles. Intra operationem cartilage of the tibial condyles was present (Type Ib, Jones et al), as indicated by the dotted line. Operative indication is for transposition of the fibula below the tibial condyles.

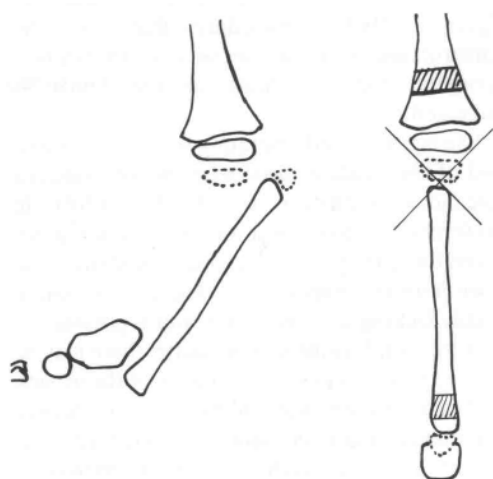


Fig. 34 The modified Brown procedure. Left, sketch of the X-ray shown in Figure 2, right. Right, the transposition of the head of the left fibula into the cartilaginous rudiment of the left tibia; fixation is by two Kirschner wires. Not shown is the Kirschner wire fixation after supracondylar shortening of the left femur and of the distal fibula before ankle fusion. Fixation of the patellar ligament to the transposed head of the fibula is done with resorbent sutures which resorb in about six weeks

In all severe cases with more than one ray deficiency of the foot, severe shortening and bowing of the tibia, the method of choice is ankle disarticulation or the formation of a modified Boyd amputation stump combined with a corrective osteotomy of the tibia (Kruger, 1971).

Contraindication for the amputation or the disarticulation is the presence of severe malformations of the upper limbs, in which case the toes are required for grasping and particularly for self-care (Fig. 39). In cases where toes must be retained we carry out lateral arthrolysis of the ankle and, if present, of the talo-calcaneo joint, disconnection of the valgus contracture and posterior transposition of the peroneal tendon(s), if necessary, after three-dimensional correction osteotomy of the tibia (Fig. 40) (Marquardt, 1981). The good results of these correlated operations have caused us recently to be more cautious with the partial foot amputation or ankle disarticulation since it has been proved that children between three years and puberty should not be amputated for psychological reasons.

I am extremely sceptical about elongations of the tibia. Blauth (1978) has published favourable results, but in his publication nothing is said about the foot and the function of the ankle joint. Figure 41, left shows the frightening result of an elongation-osteotomy of the tibia that was carried out elsewhere when the patient was an infant; the foot is in extreme valgus contracture. Disarticulation of the upper talo-calcaneo joint accompanied by the formation of a modified Syme stump was the only acceptable solution (Fig. 10, centre and right).

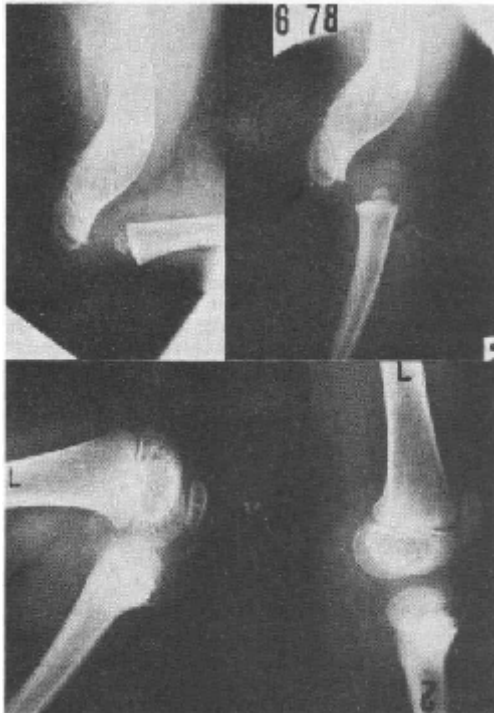


Fig. 35. Patient V.F. Top, X-ray of the right knee one year after supracondylar hyperextension and shortening osteotomy for 90° flexion contracture. The knee has only passive mobility therefore an orthoprosthesis with a knee lock is necessary. Bottom, X-ray of the left knee three years after the modified Brown procedure showing maximum active flexion and extension. The point of fixation of the patellar ligament has developed rather like a tibial tuberosity.

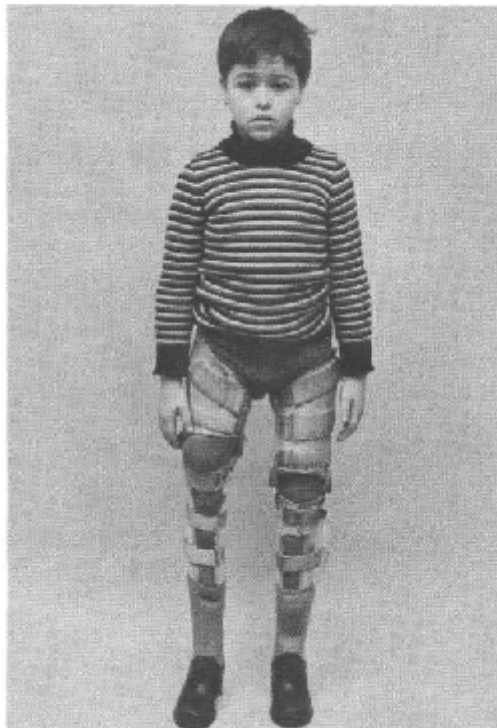


Fig. 37. Patient V.F. wears his orthoprostheses all day and is completely ambulatory. The orthoprostheses are fitted with a knee lock on the right and a free knee on the left. The thigh corset on the left side is necessary because of weak knee ligaments.

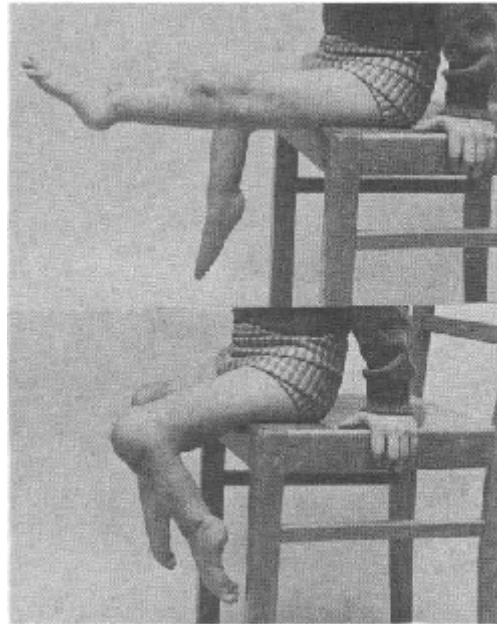


Fig. 36. Patient V.F. Active extension and flexion of the left knee three years after the modified Brown procedure.

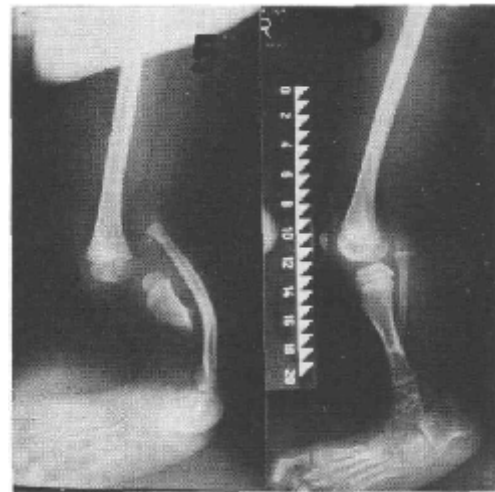


Fig. 38. Left, X-ray of the right knee, leg and foot of a four year old boy with longitudinal deficiency of tibia partial, and luxation of the knee joint caused by thalidomide. The left side is almost symmetrical. Right, two years after osteotomy of the fibula and fusion of the distal fibular fragment with the tibial rudiment, reconstruction of the knee joint and ankle fusion. There is good function in the knee. Note the adaptation of the distal fibula to tibial function. Proposed future management; partial foot amputation (modified Lisfranc) to improve the appearance and restoration to normal length by fitting an extension prosthesis.

Longitudinal deficiency femur, total or partial: PFFD

Children with a unilateral subtotal deficiency of the femur (PFFD type Aitken D) can walk without surgery by using an orthoprosthesis with a SACH foot (Fig. 42). Sooner or later, however, cosmetic problems occur due to the foot which, being at knee level, stands out in trousers or under a skirt. In addition, cosmetic and static-dynamic difficulties occur with the knee joint which, at the level of the ischial tuberosity, becomes increasingly prominent.

Surgical procedures are; the forming of a modified Syme or Boyd stump (Kruger, 1971, 1981), the tenomyoplastic Chopart stump (Marquardt, 1973) and the Borggreve (1930) and van Nes (1950) 180° rotation-osteotomy of the shank and foot with arthrodesis of the knee joint. The tenomyoplastic Chopart disarticulation and the Borggreve-van-Nes operations pre-suppose a normal ankle joint. Thus, both operations are contraindicated in the case of a combined fibular and femoral deficiency.

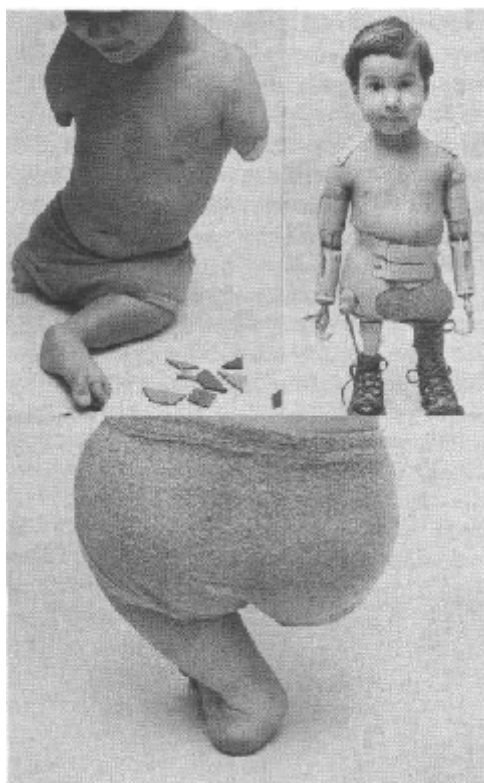


Fig. 39. Top Left, in this case (patient G.F.) of total fibular and femoral deficiency on the left side, an ankle disarticulation (modified Syme) or a Boyd amputation is absolutely contraindicated because of the high level transverse deficiencies of both arms and the phocomelia of the rudimentary right foot. (In 1975 capping procedure was carried out on the right above-elbow stump and in 1975 and 1980 on the left stump. The procedure, which in this case made use of cartilage-bone transplants from the right phocomelic extremity, is described in part II, Vol.5, No. 1.) Bottom, the increasing pes valgus makes function more difficult, this must be corrected but not amputated. Top right, the boy is ambulatory for short distances with a short Canadian hip prosthesis on the right and an orthopaedic "Innenschuh" on the left. The "Innenschuh" was unable to hold the left foot in the corrected position. The bilateral above-elbow prostheses improved the patient's balance.

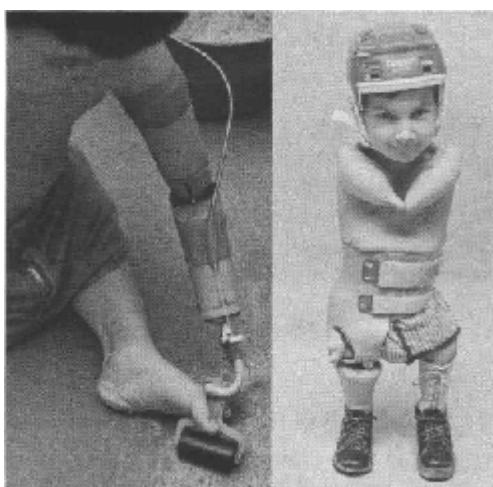


Fig. 40. Left, patient G.F. following corrective osteotomy of the tibia, lateral arthrolysis of the ankle joint and posterior transposition of the peroneal tendons to the tendon of m. tibialis posterior and to the medial ligaments; carried out in February, 1979, the function of the left foot is excellent. Right, fitting and alignment of the "Innenschuh" is now ideal. Both above-elbow stumps show considerable growth since the stump capping procedures; the stumps can be brought together which is useful for direct manipulation and for the positioning of prostheses.

can understand the transformation of the ankle joint into a knee joint by the Borggreve-van Nes operation (Fig. 43). The radically altered phenotype of the foot, which points backward, is well compensated for by the fluid walking pattern achieved which is comparable with that of the below-knee amputee. The foot is concealed by an orthoprosthesis of good cosmesis and for the swimming pool or beach a bathing prosthesis is provided.

We are still surprised that, about six months post-operatively, patients who have had this operation think in terms of knee movements while moving their ankle joint on the operated side-dorsiflexion causing knee flexion and plantarflexion causing knee extension.

Regarding bilateral PFFD type Aitken D, surgery is unnecessary for optimal fitting of orthoprostheses (Fig. 44, left). An absolute contraindication for amputation and the Borggreve-van Nes procedure is the case of bilateral PFFD combined with phocomelias or high level longitudinal and transverse deficiencies of the upper limbs (Fig. 44, right).

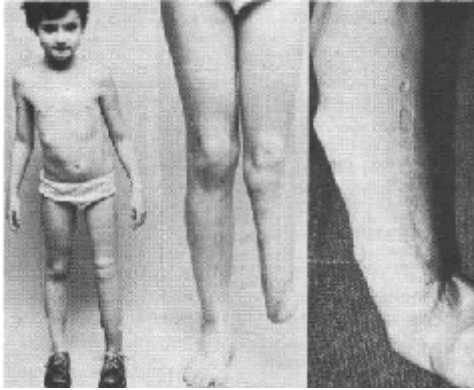


Fig. 41. Left, right leg of a four year old child with longitudinal deficiency fibula and ray IV and V total showing extreme pes valgus after elongation of the tibia (see text). Centre, the right leg after distal tibial resection (modified Syme). Right, the patient fitted with a modified PTS prosthesis.

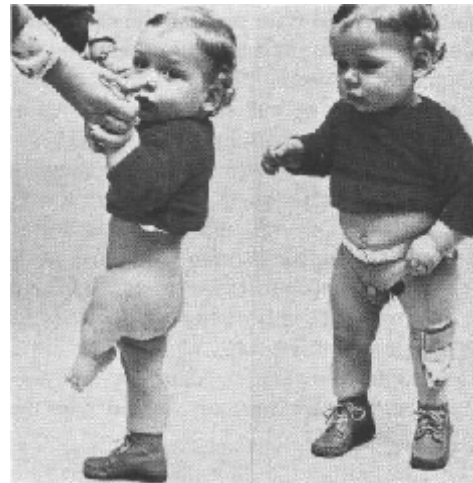


Fig. 42. Left, fourteen month old child with unilateral PFFD type Aitken D. Right, the child ambulatory with an orthoprosthesis without knee joint. SACH foot fitted.

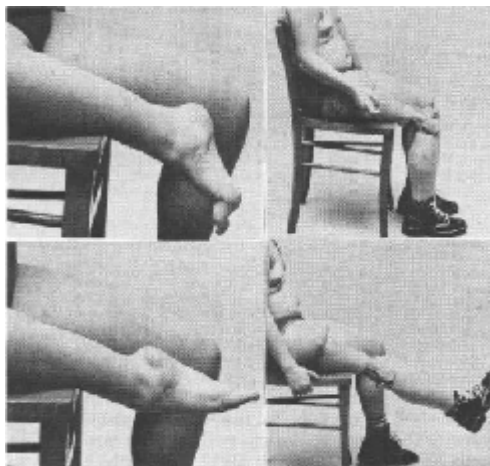


Fig. 43 Transformation of the ankle joint into a knee joint by the Borggreve-van Nes procedure. Top, dorsiflexion of the ankle joints effects knee flexion. Bottom, plantarflexion effects knee extension.

I do not wish, in this contribution, to go into detail regarding the operative treatment of PFFD type Aitken A, B and C; it is a matter of aligning the neck of the femur by resection of the subtrochanteric pseudarthrosis and taking advantage of the knee joint in the best way possible.

Transverse lower limb deficiencies require surgical intervention only if there is a threatened perforation by the bone through the skin of the below-knee stump (American Academy of Orthopaedic Surgeons, 1981).

I shall report in our next Congress about surgical stump elongations, for example, the elongation of an ultra-short above-knee stump to provide a reasonable length for fitting an above-knee prosthesis. The majority of the examples discussed of operative interventions, particularly for the lower limb, can provide maximum benefit for the patient only when supported by appropriate prosthetic care.

Operation, prosthetic technique, physiotherapy and ergotherapy are bound into the same

rehabilitative concept with the goal of reaching, or at least facilitating, mastery of the patient's everyday life, integration into school, occupation and participation in social life, including sports and leisure activities.

I have shown only some small elements; a lot of work still lies ahead. In preparing ourselves for this responsible task, Knud Jansen has pointed out that the way is by multidisciplinary co-operation.

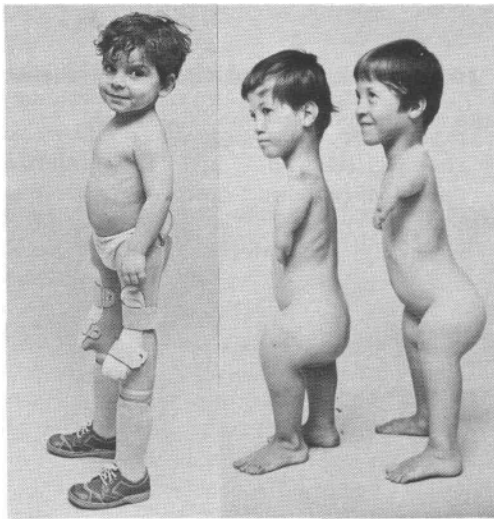


Fig. 44. Left, child with PFFD type Aitken D bilateral and normal upper limbs. No surgery is necessary for optimal fitting of orthoprostheses. The axis of the knee joints should be 3-4 cm higher than shown in the photograph. Right, twins with severe longitudinal deficiencies of both upper and lower limbs caused by thalidomide. In these cases of bilateral PFFD type Aitken D, amputations or Borggreve-van Nes procedures are absolutely contraindicated. Both children are ambulatory for short distances. For longer distances they use electrically driven wheelchairs.

Acknowledgements

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