

THE KNUD JANSEN LECTURE

The operative treatment of congenital limb malformation—part III

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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 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. 1, left).

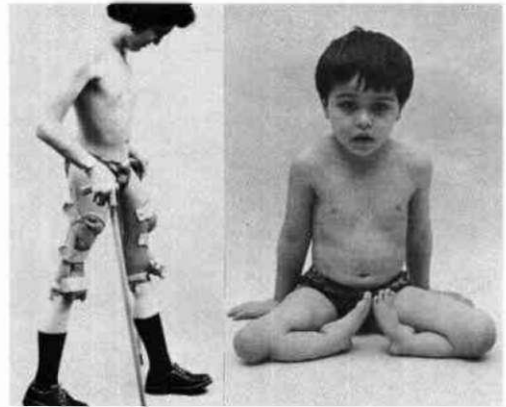


Fig. 1. Left, patient with longitudinal deficiency tibia, total bilateral, right coxa vara, PFD type Aitken A left with sub-trochanteric pseudarthrosis; longitudinal deficiencies of metacarpal and phalanges total. The left hip was reconstructed and both knees and ankles fused. Patient is ambulatory (for 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.

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

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essential as the proximal epiphysis 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. 1, right and Figs. 2-6). 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. 7).

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

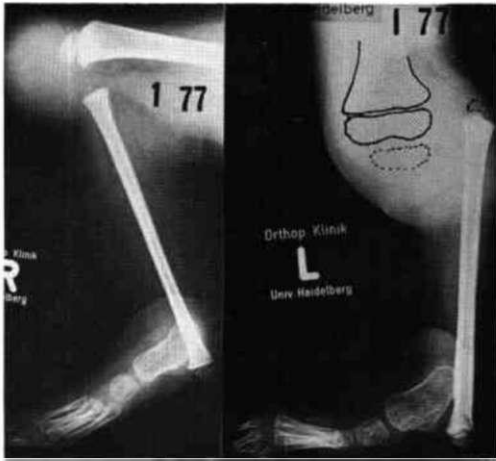


Fig. 2. Left, patient V.F. The X-ray of the right leg shows a longitudinal deficiency tibia, total. Type 1a (Jones et al, 1978). The Brown procedure is contraindicated because of the patient's age and the hypoplastic distal femur. Indication is for knee and ankle fusion or, as done, supracondylar osteotomy 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 1b, Jones et al), as indicated by the dotted line. Operative indication is for transposition of the fibula below the tibial condyles.

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.

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. 8). 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. 9) (Marquardt, 1981). The good results of these correlated operations have caused us recently to be more cautious with the partial foot

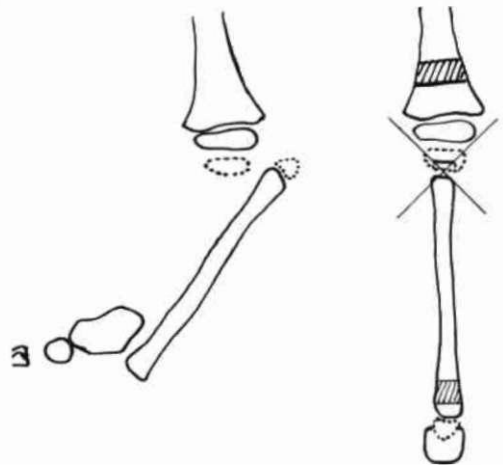


Fig. 3. The modified Brown procedure. Left, sketch of the X-ray shown in Figure 2, right. Right, the transposition of the head of the left fibula into the cartilaginous rudiment of the left tibia; fixation is by two Kirschner wires. Not shown is the Kirschner wire fixation after supracondylar shortening 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.

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 10, 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. 4. 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. 5. Patient V.F. Active extension and flexion of the left knee three years after the modified Brown procedure.

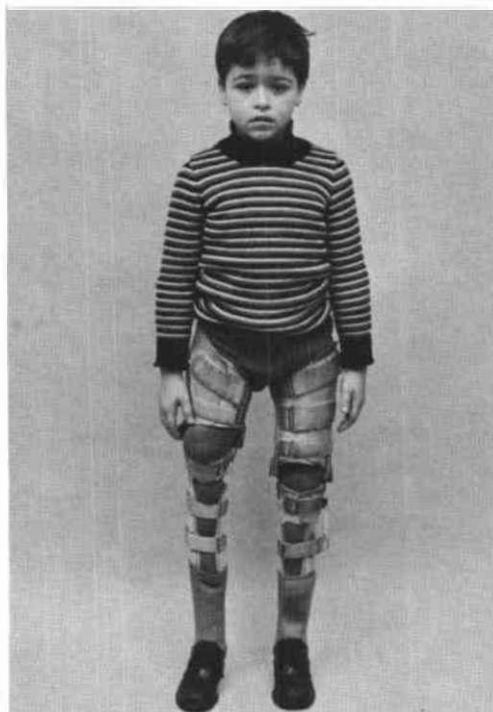


Fig. 6. Patient V.F. wears his orthoprosthesis all day and is completely ambulatory. The orthoprosthesis are fitted with a knee lock on the right and a free knee on the left. The thigh corset on the left side is necessary because of weak knee ligaments.

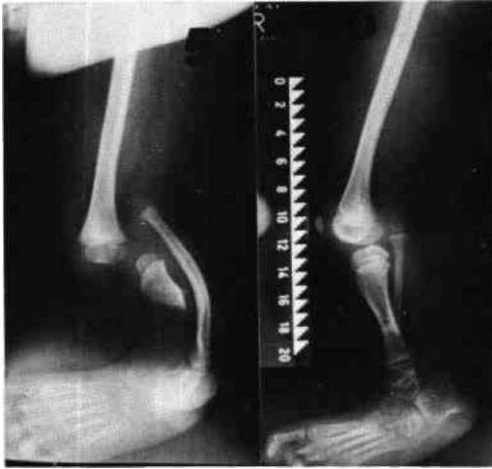


Fig. 7. Left, X-ray of the right knee, leg and foot of a four year old boy with longitudinal deficiency of tibia partial, and luxation of the knee joint caused by thalidomide. The left side is almost symmetrical. Right, two years after osteotomy of the fibula and fusion of the distal fibular fragment 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. 11). 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. 8. 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.

The pros and cons of these operations must be carefully discussed with the parents and the patient with the help of other patients who have undergone surgery and with the aid of photographs and films. Consultation with patients who have already had the operation is most valuable so that the patient and parents can understand the transformation of the ankle joint into a knee joint by the Borggreve-van Nes operation (Fig. 12). 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.



Fig. 9. 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.

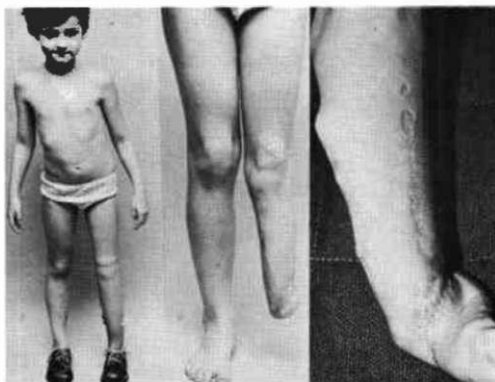


Fig. 10. Left, right leg of a four year old child with longitudinal deficiency fibula and ray IV and V total showing extreme pes valgus after elongation of the tibia (see text). Centre, the right leg after distarticulation of the ankle joint (modified Syme). Right, the patient fitted with a modified PTS prosthesis.

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. 13, left). An absolute contraindication for amputation and the Borggreve-van Nes procedure is the case of

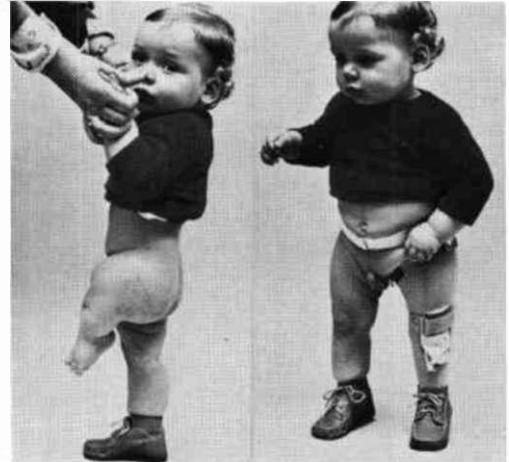


Fig. 11. Left, fourteen month old child with unilateral PFFD type Aitken D. Right, the child ambulatory with an orthoprosthesis without knee joint. SACH foot fitted.

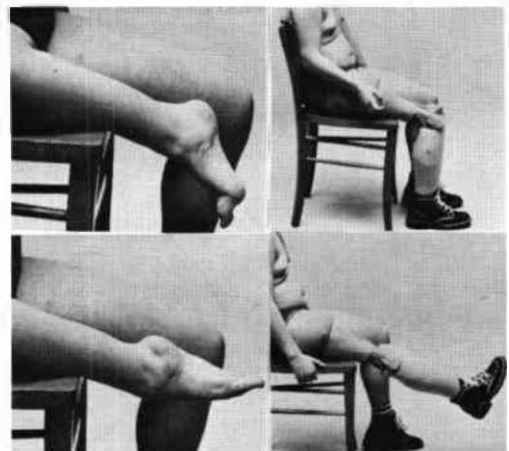


Fig. 12. Transformation of the ankle joint into a knee joint by the Borggreve-van Nes procedure. Top, dorsiflexion of the ankle joints effects knee flexion. Bottom, plantarflexion effects knee extension.

bilateral PFFD combined with phocomelias or high level longitudinal and transverse deficiencies of the upper limbs (Fig. 13, right).

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 sub-trochanteric 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

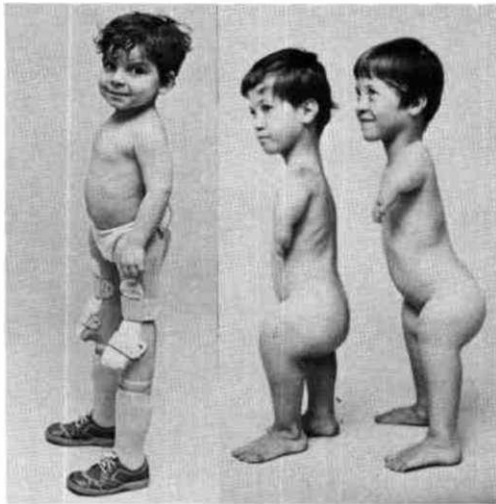


Fig. 13. Left, child with PFFD type Aitken D bilateral and normal upper limbs. No surgery is necessary for optimal fitting of orthoprostheses. The axis of the knee joints should be 3-4 cm higher than shown in the photograph. Right, twins with severe longitudinal deficiencies of both upper and lower limbs 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.

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.

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