

The care of the limb deficient child in North Africa

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Abstract

The surgical and prosthetic treatment of longitudinal lower limb deficiency is described and discussed, in the light of cultural and social requirements. Those with upper limb deficiencies are not fitted with prostheses.

Introduction

Cases of congenital limb deficiency have always been a drama for the family, a handicap for the patient and a heavy burden for the society. The Kassab Institute has devoted much attention to these cases which are seen as being very important.

Our therapeutic attitude, which was initially conservative (due to the social and cultural context and to fitting difficulties) is becoming less restricted; the main objective of treatment is to allow the child to carry out the activities of his age group.

The purpose of this paper is to report our findings concerning the care and the course of the treatment of children suffering mainly from a failure of formation of the lower limb.

During the period between 1970-1990, 74 children (40 boys and 34 girls) received treatment at the Tunis orthopaedic Institute M. T. Kassab.

There were 88 affected limbs, 77 longitudinal deficiencies and 11 transverse deficiencies (12 patients with 16 associated upper limb anomalies).

Fibular deficiency (2 groups)

First group

Some 18 children (21 limbs) were seen at the Institute at an average age of six months (varying from 11 days to 11 months).

The surgical treatment was carried out at the age of 12 months in the following way:

— 14 patients had a foot correction and realignment for marked talipes equino-valgus (resection of fibrous lateral band, lengthening of achilles and peroneal tendons and a posterior capsulotomy).

— 8 had an osteotomy of the tibia for anterior bowing of this bone. After this surgery, walking was possible using an orthosis which contained the corrected foot and compensated for the shortening (Fig. 1).

During the course of the treatment, there was an increase of the shortening and the orthosis was given up by 11 who preferred walking on their toes without an orthosis. The equinus compensates for the shortening and the foot remains in axial alignment.

In three cases the equinus of the foot could not compensate for the shortening and a lengthening of the tibia by the Wagner procedure was performed. Four children were fitted with a definitive extension or ortho-prosthesis because of severe knee instability.

Second group

Fourteen older children were first seen between the ages of three and 16 years. Nine of them walk on their toes with moderate talipes

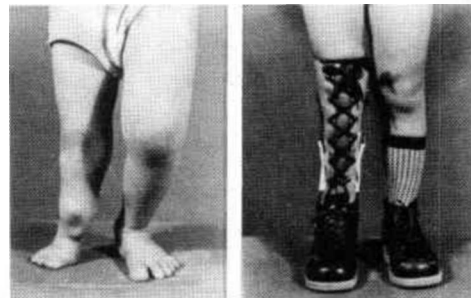


Fig. 1. A child, four years old with fibular deficiency, operated on and fitted with simple orthosis.

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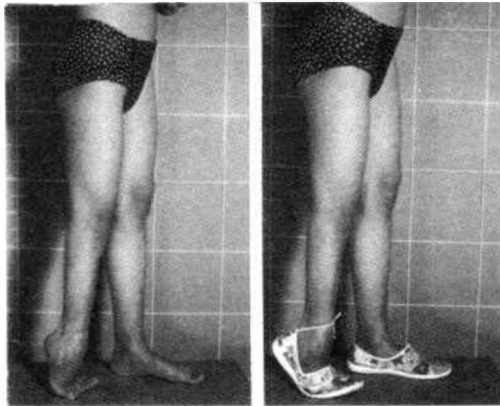


Fig. 2. A girl, 15 years old with fibular deficiency, walking without orthosis in a digitigrade way.

equino-valgus. They were treated conservatively as their walk was balanced, the knee stable and the shortening compensated by the equinus (Fig. 2). This approach is always accepted well by the parents.

Five walk with a pronounced limp due to shortening, knee instability, and in two cases hip dislocation. An ortho-prosthesis was fitted to four straight away and to the fifth after failure of surgical equalisation.

Tibial deficiency

Total absence – Type 1 (Table 1)

There were 10 patients who presented with a total of 14 limbs in this group.

Disarticulation of the knee followed by prosthetic fitting was always carried out at an average age of three years because of the severe knee flexion and foot deformity (Fig. 3).

Distal 2/3 partial absence – Type 2 (Table 1)

There were 10 patients who presented with a total of 11 limbs in this group.

Four patients had realignment surgery,

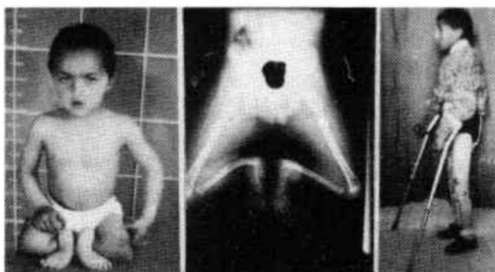


Fig. 3. A case of bilateral tibial deficiency fitted with prosthesis after knee disarticulation.

Table 1. Classification of tibial deficiency:

- Type 1: total absence
- Type 2: distal 2/3 partial absence
- Type 3: distal 1/3 partial absence

Type 1	Type 2	Type 3
14	10	4

consisting of fixation of the foot under the fibula, followed by fusion of the fibula to the tibia when about two years old. An orthosis maintains the corrected alignment of the leg and foot and compensates for the shortening (Fig. 4). Three of these patients had leg lengthening later at the age of 10 using Wagner or Orthofix devices.

Six patients had severe foot deformity and marked fixed flexion of the knee. Despite all attempts to reconstruct, three had to have an amputation and three had knee disarticulation, followed by prosthetic fitting.

Distal 1/3 partial absence – Type 3 (Table 1)

There were 4 patients who presented with a total of 4 limbs in this group.

Table 2. Asmutz classification of proximal femoral deficiency

I	II	III	IV	V
6		4	3	1

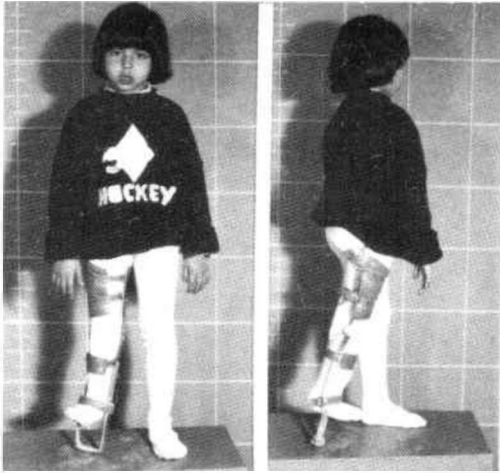


Fig. 4. A girl, five years old with tibial deficiency Type II, operated on and fitted with orthosis with compensation for shortening.

All presented with a stable knee and only minor foot deformity. All were fitted with an orthosis. Two have been lengthened and the others await this procedure.

Femoral deficiency

Amstutz – Type I (Table 2)

There were 6 patients who presented with a total of 6 limbs in this group.

Five patients underwent a correction of the coxa vara by valgus osteotomy of the femur. An extension or orthoprosthesis compensating for the marked shortening produced a balanced walk. In this group three children had a previous femoral equalisation at the age of 12 (femoral lengthening and contralateral epiphysiodesis).

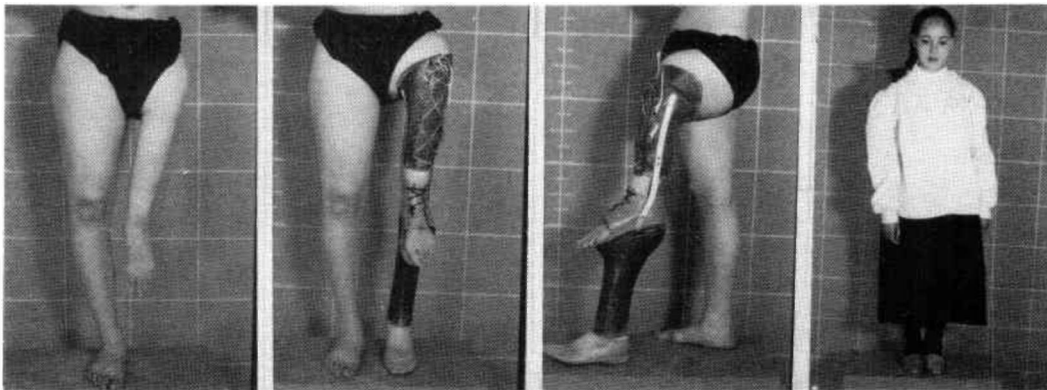


Fig. 5. A girl, 15 years old with partial femoral deficiency Type IV, fitted with ortho-prosthesis with which she is independent.

Amstutz – Types III, IV and V (Table 2)

There were 5 patients who presented with a total of 8 limbs in this group.

These were treated conservatively with orthoprotheses because of the marked femoral hypoplasia (Fig. 5).

Transverse deficiency

Nine patients (11 limbs) were provided with prostheses whose type depended on the age, the level of loss and the social environment.

Results and discussion

Limb deformities are relatively rare in our country (less than 3 in 1,000 births), which corresponds to the classical data (Thevenin *et al.*, 1988; Schoenecker *et al.*, 1989; Briard and Boullegue, 1978). Our patients are independent despite the severity of the limb lesions and the simplicity of our therapeutic approach.

Ten are doing sedentary jobs, 10 are working in agriculture, 30 are still attending school and 11 could not find employment.

Fibular deficiency

No matter what degree of deformity and the age of the patient at first consultation, surgical intervention remains simple. Correction of the valgus is maintained and walking on the toes in equinus is well tolerated in our rural environment.

Tibial deficiency

The severe deformities associated with Type 1 cases and with those of Type 2 with severe foot deformity and knee laxity led to early amputation (Schoenecker *et al.*, 1989; Epps and Schneider, 1989; Frantz and O'Rahilly, 1961;

Bardot and Bouyala, 1978). Fitting bilateral cases remains difficult and the prostheses are not always tolerated, which reduces the patient's activity and heightens the psychological impact of amputation or disarticulation on the Tunisian patient personality.

Those with a stable knee whose foot can be preserved (some Type 2 and all Type 3) present a problem of length inequality. Contralateral tibial epiphysiodesis may avoid some complications.

Femoral deficiency

Correction of the coxa vara restores the architecture of the hip and provides femoral lengthening of 2–3 cm. The percentage of shortening remains constant compared to the length of the contralateral femur (Carlioz, 1978; Thevenin *et al.*, 1988). Final equalisation is performed at the age of 12 to 13 years to avoid multiple surgery. Rotationplasty has not been performed as it is always refused by the parents despite hip instability, malrotation with muscular inadequacy and even complete absence of femur (Amstutz Type 1). In these cases an ortho-prosthesis is accepted as the final treatment. No amputation has been carried out in this group of patients. Four adults prefer to use a wheelchair.

Upper limb deficiency associated with lower limb deficiency

Twelve patients have 16 upper limb deficiencies. They adapt easily to their handicap and are not fitted with prostheses.

Conclusion

Despite the complexity of these deformities the results remain satisfactory, being influenced

by the severity of the shortening in the unilateral cases, the position and stability of the foot, knee and hip and deformity of the segments. We think the foot and leg should be preserved, maintaining the equinus position allows the use of an orthosis which is distal to the knee and allows better growth of the affected segment.

Amputation or disarticulation should only be used as an inferior solution in our social and economic environment.

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