THE NATURAL HISTORY AND MANAGEMENT OF
CONGENITAL SHORT TIBIA WITH DYSPLASIA OR
ABSENCE OF THE FIBULA

A PRELIMINARY REPORT

D. HOOTNICK, N. A. BOYD, J. A. FIXSEN AND G. C. LLOYD-ROBERTS, LONDON, ENGLAND

From the Orthopaedic Department of The Hospital for Sick Children, Great Ormond Street, London

Forty-three patients with unilateral congenital short tibia with partial or complete absence of the fibula are reviewed. The factors influencing the degree of leg shortening at maturity are considered. Serial radiographic measurements of leg length in fourteen patients covering an average observation period of 9-3 years support the hypothesis that the relative difference in growth between the two limbs remains remarkably constant. By estimating the percentage difference between the normal and abnormal leg lengths on the first measurable radiograph it is therefore possible to predict the likely shortening at maturity. This method of prediction allows the surgeon to make the decision to proceed to Syme's amputation or to the use of an extension prosthesis at about one year of age when the child starts to walk.

Rational management of an infant with unilateral congenital shortening of the tibia with complete or partial absence of the fibula depends upon our ability to predict the likely discrepancy of leg length at maturity (Figs. 1 and 2). The confidence of the surgeon in electing either to amputate early in childhood or to prescribe an extension prosthesis, accepting, modifying or correcting the deformity of the foot, is directly related to the accuracy of this prediction. We will consider the features upon which prognosis may depend and describe a method of prediction, which seems to be sufficiently accurate for our purpose.

Publications on this topic are scanty and somewhat confusing. Coventry and Johnson (1952) related the prognosis directly to the degree of morphological abnormality of the limb as a whole, while Kruger and Talbott (1961) described an inverse relationship between the degree of maldevelopment of the foot and ultimate shortening: the better the foot the shorter the leg. These propositions will be assessed later.

THE BACKGROUND TO THE INVESTIGATION

The policy adopted for many years at the Hospital for Sick Children has been to decide whether to proceed to Syme's amputation or management by an extension prosthesis at about one year of age, so that walking is established in a manner that is likely to be permanent. We believe that when amputation seems to be preferable it is less distressing to the child and parents if done at this early age rather than some years later, when the prosthesis may be rejected for cosmetic or other reasons.

When the overall leg discrepancy has been between 3-7 and 5 centimetres at one year amputation has been advised, for we believe that the ultimate shortening will be within the range of 10 to 12-5 centimetres. Discrepancy of this order is beyond redemption by shortening of the normal side within acceptable limits. Tibial lengthening in this condition has, in our hands, proved a particularly difficult undertaking, with delayed union, supplementary grafting and some loss of final correction as the penalty. After both lengthening and shortening a final difference of more than 5 centimetres is probable, retaining a foot with variable deformity and rigidity, to be accommodated by a specially made and often ugly raised shoe.

We have not, in general, attempted correction of the deformity of the foot when this is preserved, for the prosthesis may be suitably adapted.

The purpose of this review is to test the validity of this policy and to re-examine some of the clinical features of this malformation in so far as they may help us to predict the final outcome with greater accuracy at an early age.

CLINICAL MATERIAL

We have studied the records of forty-three patients with congenital shortening of the leg in whom there was a tibial component. Those selected for study fulfilled the following criteria. The defect was strictly unilateral. Measurements of the initial and final shortening from
proximal to distal tibial epiphysis were calculated from scanograms or in some very young children from radiographs showing the two tibiae on one plate. Anterior bowing of the tibia is commonly present in this condition but the vertical height of the tibia is the same when measured on antero-posterior and lateral scanograms. Therefore, standard antero-posterior scanograms were used for our calculations. Syme's amputation did not affect the result, for the foot was always excluded from the measurement. Clinical estimates were ignored as being unreliable and erratic. The ages of first and last assessment, were, therefore, determined by the availability of acceptable radiographs. Most had associated femoral shortening, but we rejected those in whom the femoral dysplasia was of a degree greater than simple shortening—that is, those with coxa vara, proximal deficiency, etc. In no patient was the femoral component greater than the tibial.

Unfortunately, many patients lacked acceptable early radiographs or were unavailable for final examination. There remained fourteen patients, fully documented and personally examined, whose ages at final examination ranged from eight to twenty years, with a mean of 12·5 years and with satisfactory radiographs covering an average observation period of 9·3 years.

Leg lengthening had been performed in two of these, which were included, with measurements adjusted to exclude the gain obtained, as estimated from scanograms before and after the lengthening. Twelve patients had some degree of associated femoral shortening, which must be recognised as an important component in the syndrome of congenital shortening of the tibia.

The extremes of final shortening were widely spaced. In one patient of ten years the tibia was 17 centimetres short, whereas in another of thirteen years the deficiency was only 5 centimetres. Mean measure-

![Fig. 1](image1.png)

**Fig. 1**
Anterior and lateral photographs of a child aged 11½ months with a congenital short tibia on the right. Note the foot deformity, the tibial kyphosis and the skin dimple.

![Fig. 2](image2.png)

**Fig. 2**
Anterior and lateral photographs of a patient with congenital shortening and bowing of the tibia and a dimple on the right.

ments are, therefore, of no significance because of the age and anatomical differences between patients.

**FACTORS INFLUENCING PROGNOSIS**

The foot—It has been suggested (Kruger and Talbott 1961) that shortening is inversely related to the state of preservation of the foot—the more normal the foot the greater the shortening. Four grades of progressive malformation were described, based on the number of metatarsal bones remaining. All four grades were seen among our fourteen patients, but there was no apparent correlation.

In thirty-six of our total of forty-three patients this aspect could be analysed because early radiographs were not necessary. Our findings show the opposite trend to that suggested by Kruger and Talbott, and are in accord with those of Coventry and Johnson (1952) (Table 1). Those with fewer metatarsals tended to have the greater shortening, but the range in any one group was large, rendering this method of prediction unreliable in the individual patient.
The fibula—The fibula was absent in eleven patients and present but abnormal in three. Although in one, with approximately four-fifths of normal development, shortening was only 4.9 centimetres at the age of four-
teen years, two with half and two-thirds development were 10 centimetres and 10.5 centimetres at fourteen and sixteen years respectively. These are within the range observed in complete absence of the fibula, and correlation of predicted limb length with the degree of preservation of the fibula is poor.

Corrective osteotomy of the tibia was necessary in only two patients to aid fitting the prosthesis. The angulation tends to correct itself spontaneously with time (Farmer and Laurin 1960).

Femoral shortening—Although a femoral component is frequently associated with shortening of the tibia

Tibial kyphosis—Anterior tibial bowing was somewhat arbitrarily estimated from the first radiographs as absent (5), moderate (4) or severe (5). There was no correlation with the final outcome, or with foot deformity.
(twelve of the fourteen patients), the wide variation in this contribution to the final discrepancy (0·5 centimetre to 7·7 centimetres at ten years) prevents its recognition alone from becoming a useful guide to the prognosis.

A METHOD OF PREDICTING SHORTENING AT MATURITY

It is evident that the factors already analysed, whether considered singly or in combination, do not enable us to predict the outcome with sufficient confidence for a decision to be made for or against amputation before walking begins.

We therefore investigated the possibility that the relative difference between the limbs remained the same throughout growth. Ring (1959), working at this hospital, demonstrated that the relative difference remained constant in patients with unilateral congenital short femurs. This proposition had been suggested by Aitken (1974) and more recently in relation to congenital short tibia by Westin, Sakai and Wood (1976).

The importance of the femoral component has already been emphasised, and consequently for accurate prediction it is essential to include the femur in the calculation.

Our method first estimates the percentage difference between the normal and abnormal femur and tibia from the first measurable radiograph. The height of the patient at maturity is then estimated from parental heights. This projected height and the probable normal femoral and tibial lengths can be readily obtained from available growth charts. Deduction of the initial percentage difference from the normal projected leg length gives an estimate of the final discrepancy.

Example

Case 1—The patient's first measurable radiograph was at six months, when the shortening was 3·7 centimetres. The femur was 94 per cent of normal length and the tibia only 76 per cent. The father was 180 centimetres tall and the mother 135 centimetres. From growth charts prepared by Tanner, Whitehouse and Takaishi (1966) we found that the father's height lay on the 90th percentile of the population and the mother's on the 50th percentile. Taking an average of the 70th percentile, we estimated the patient's height at maturity, as 167 centimetres. (The estimation of mature height in a patient under the age of one year is difficult, but the method described using percentiles of the father's and mother's height is the most accurate available at this age (Whitehouse 1976).) The tables of Anderson, Green and Messner (1963) predict that an individual of this height will have a femur of 45·3 centimetres and a tibia of 36 centimetres.

If we now deduct the initial percentage deficiency from femoral and tibial length we can predict a femur of 42·6 centimetres and tibia of 27·4 centimetres. The calculated shortening at maturity is, therefore, 11·3 centimetres.

The patient was re-examined at the age of fourteen years and six months. His height was 164·5 centimetres (an error of 2·5 centimetres), and the total shortening 9·8 centimetres which represents an error of 1·5 centimetres.

There were four additional patients over fourteen at the final examination. The percentage differences remained remarkably constant (Table II).

Furthermore, the same pattern is developing in the remaining nine patients between eight and fourteen years old (Table III). In addition three patients not
previously included, aged three-and-a-half to five, show a similar trend. In spite of their youth they are significant because at three-and-a-half years the normal tibia has grown to half its final length.

We appreciate that the method used is somewhat tedious, but it is probable that by direct measurement of the leg bones of the parent of similar sex and deducting the percentage deficit in the child we would obtain a forecast sufficiently accurate for clinical purposes.

**APPLICATION TO MANAGEMENT**

We do not intend to discuss management in detail, but merely to state our views on the indications for and the technique of amputation. Our experience with a much larger number of patients in whom amputation has been performed or the foot preserved supports certain conclusions.

If the predicted shortening is less than 8.7 centimetres, conservation would seem appropriate. Between 8.7 centimetres and 15 centimetres amputation is indicated, for such a discrepancy is beyond surgical redemption and an extension prosthesis less desirable. If shortening is more than 15 centimetres a retained foot has some advantage as a means of lengthening the below-knee component, provided it can be placed in full equinus without valgus. If valgus cannot be corrected due, for example, to the shape of the lower tibial epiphysis (Figs. 3 and 4), the prosthesis becomes difficult to fit and cumbersome so that amputation is preferable.

Our views on amputation are therefore very similar to those of Farmer and Laurin (1960) and Wood, Zlotsky and Westin (1965).

A modified Syme’s amputation is recommended. The lower tibial epiphysis is preserved, the medial malleolus being cut at the level of the tibial articular cartilage. There is one technical problem peculiar to this condition. Tibial kyphosis will tend to displace the heel flap backwards away from the weight-bearing area. If the kyphosis is moderate it is enough to secure the flap to the tibia by a Kirschner wire. If, however, kyphosis is severe it is preferable to correct the angulation by an osteotomy at the time of amputation securing flap and tibial osteotomy with the same wire (Fig. 5).

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**REFERENCES**


