Congenital Absence of Tibia

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Abstract

Congenital absence of tibia is a rare anomaly. We report a case who presented at the age of 3 years with absence of tibia right side with associated anomalies and was managed by reconstruction of the knee and ankle joints by transfer of fibula

Keywords

Hemimelia, Equinus

Introduction

Tibial hemimelia is an extremely rare entity. The condition must be distinguished from the commonly occurring hypoplasia or absence of fibula. We present a case with a congenital absence of tibia, who at the time of presentation had grossly deformed and functionally useless leg. The purpose of this presentation is to assess the various modalities of management as written in literature and compare them with our experience.

Case Report

A 3 year old boy (R.S.) came to us with history of multiple deformities involving the right knee, ankle, foot, both hands and scrotum since birth. On examination the child had severe flexion contracture of the right knee, medially deviated leg, prominence of fibular head posterolateral riding over the lateral femoral condyle. Active extension of the knees was possible upto 45° with full flexion at knee joint. The right leg was hypoplastic and severely shortened. The right foot was fully adducted facing the perineum with the ankle in equinus and only a single bone (fibula) was palpable in the leg. Prominent lower end of the fibula was palpable on the lateral side of the ankle. Associated anomalies included hypoplasia of right thumb, congenital hydrocele(right) and a mild hyperkinetic behaviour (Fig. 1).

Fig 1. Showing adducted foot with ankle in equinus with hypoplasia of right thumb and hydrocele.

Investigations done included routine haemogram, serum chemistry, B. Sugar, chest x-ray, x-ray spine. All these investigations were within normal limits. X-ray of the involved leg showed a single bone (fibula) overriding the femur proximally and calcaneum distally (Fig. 2).
The treatment followed in the case was reconstruction of the knee by proximal end transfer of fibula under the femoral condyles (Stage-I). After 6 weeks lower end of the fibula was fixed in the centre of talus (rudimentary) and calcaneum and stabilized using a K-wire through calcaneum and talus into the fibula and foot was held in equinus with no adduction or abduction (Stage-II). The equinuous is thought to be advantageous as it adds to the length of the shortened leg.

The patient was reexamined after 1 month and the appearance and alignment of both knee and ankle was reasonably good clinically (Fig. 3) and radiologically (Fig. 4) but the long term results are still awaited.

Discussion

The case of our patient who had congenital total absence of tibia is extremely rare. Only 250 cases have been reported in the literature till date (1). It is usually associated with other anomalies like synclinodactyly, anomalies of the genitourinary tract, mental retardation etc. (2) which was true in our case also.

The various modalities of treatment advocated are:

a) Disarticulation at knee (3) b) Amputation through leg and arthrodesis of knee (4) c) Transfer of fibula to knee (5) d) Orthodoxies of fibula to femur and ankle (6)

We followed the treatment protocol as advocated by Brown (5) as the child being only around 3 years old had the potential of fibula taking over the function of tibia with persistent use and weight bearings, instead of being given an artificial limb after disarticulation through the knee. The patient was made ambulatory with a long leg caliper.

References

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