LENGTHENING OF THE LOWER LIMBS AND CORRECTION OF LUMBAR HYPERLORDOSIS IN ACHONDROPLASIA

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ABSTRACT

This paper describes the technical development as well as results obtained with the method of bone lengthening developed by the author and used over the past fifteen years for the treatment of short-statured conditions not susceptible to hormonal treatment.

MATERIAL AND METHODS

One hundred and seventy-two patients have been treated since 1970 to this date. The patients' ages range between 4 and 30 years with a mean of 14 years. Bone lengthening was performed to address shortness of stature caused by the osteochondrodysplasias (Achondroplasia), syndromes of hormonal deficit refractory to treatment with growth hormone, Turner's syndrome, etc. One hundred and thirty-three patients had achondroplasia within this series. Bone lengthening was practiced on 596 bone segments, of which 344 were tibias and 252 femurs.

This series includes cases treated initially with the original method, which required osteosynthesis as a second stage of treatment, as well as cases treated with the present method. This paper restricts itself to the evaluation of cases treated with the present method. It includes 104 patients, with treatment performed on 208 tibias and 156 femurs.

SURGICAL PROTOCOL

Patients are evaluated in the Unit of Growth Disorders by a team consisting of a pediatrician, an endocrinologist, an orthopaedic surgeon, a geneticist, a specialist in rehabilitation, and an orthotist. They are subjected to clinical and radiological evaluation, and numerous tests with the purpose of establishing a complete etiological diagnosis. A treatment
plan is then elaborated for each individual patient based on this evaluation. If feasible, the endocrinologist will prescribe hormonal management. For cases where this is not an option, and whenever the general condition of the patient permits (age, physical and psychological make-up, etc.), surgical treatment is recommended. The presence of hydrocephalus or symptomatic spinal stenosis specifically is ruled out in achondroplasts.

For this purpose, the patients are examined by the neurosurgeon and CT or MRI scan are prescribed, as well as an electromyogram and sonogram as indicated. If the above tests rule out neurological pathology and if the patient as well as his close relatives are in agreement with our surgical management program, the treatment is started.

Leg lengthening is not recommended for patients younger than 7 years of age since this might cause significant setbacks in schooling as well as to psychological make-up and interfere with the development of early interpersonal relationships with peers as the early phases of treatment promote a diminished body image (wheelchair). Also, the quality of regenerated bone appears to be better between 7 and 11 years of age. The lengthening is carried out bilaterally on the same segment. It starts with the tibiae and continues with the femora in a second stage. A prudent amount of time is allowed between the lengthening of each set of segments to permit the patient to regain his functional gait between each intervention. We have ruled against performing either cross lengthening (femur on one side, tibia on the other) or linear lengthening (femur and tibia on the same side). In either instance, if the treatment had to be discontinued because of a severe complication, the patient would be left with a serious asymmetry or disharmony for which compensation would be very difficult. Furthermore, the lengthening of the femur and tibia on the same side creates excessive stress on the soft tissue structures of that limb (muscles, vessels, nerves, etc.) and also obligates the patient to wear a shoe lift of progressive thickness to compensate for the gradually increasing length of the opposite limb. With the method of parallel lengthening, the treatment can be discontinued at any time if forced by the circumstances.

SURGICAL TECHNIQUE

Tibial lengthening

First, the distal tibio-fibular joint is internally fixed with a Palmer nail to prevent the development of progressive valgus deformity of the ankle during lengthening (Fig. 1a,b,c,d). Next, two parallel positioned Schanz screws are inserted from medial to lateral through the proximal and distal tibial metaphyses. The most proximal screw is inserted just distal to the proximal tibial physis. The double screw proximally and distally creates a greater stability of the segments and minimizes the tendency to angular deviation of the segments and minimizes the tendency to angular deviation of the segments during lengthening (in varus, valgus, or recurvatum).

The excisional osteotomy of the fibula should be at least 1.5 cm.