

FIBULAR PACHYDYSOSTOSIS : AN ATYPICAL CASE

J. C. ABRIL, A. DÍAZ, T. EPELDEGUI

Pachydysostosis of the fibula is an isolated deformity of the fibula with bowing and lengthening. Only one report has appeared in the literature, describing four lesions which underwent spontaneous regression during early childhood ; no therapy was recommended.

We report a 12-year-old boy with this type of deformity, who developed progressive angulation and deformation of the ankle joint. An osteotomy was necessary to realign the fibula. One year later the patient was free of symptoms.

Keywords : fibular pachydysostosis ; fibula bowing ; osteotomy.

Mots-clés : pachydysostose ; péroné ; incurvation ; ostéotomie.

INTRODUCTION

Pachydysostosis of the fibula is a new entity described by Maroteaux *et al.* in 1991 (5). All four cases that have been described presented an isolated deformity of the fibula with bowing and lengthening. All were congenital, and underwent spontaneous regression during early childhood. No therapy was recommended, and all of them ended up with a slight residual deformity.

We report a 12-year-old boy with this type of deformity.

He complained of pain and tenderness in his right calf, without any apparent clinical deformity. During the previous year, progressive angulation and destruction of the ankle joint were observed. Surgical treatment with osteotomy was required to realign the fibula.

CASE REPORT

A 12-year-old boy complained of pain in the right calf during the previous year. There was no relation to trauma or sports. Six months later, the pain had increased and moved into the ankle area, and the patient limped. No growth hormone (GH) had been previously administered. Clinical examination showed calf atrophy, although the fibula was not palpable. There was a 10° limitation active dorsiflexion of the ankle. Blood analyses were normal. X rays showed a deformity of the fibula with posterior bowing and increased length and thickness (fig. 1).

No other skeletal anomaly in the other bones was observed by x ray. MRI ruled out associated muscular or vascular anomalies in the affected leg. No signs of neurofibromatosis were found.

A shortening osteotomy was performed with resection of 2.5 cm of the distal third of the fibula ; a cast was applied for two weeks, after which progressive weight bearing was permitted. Six months after surgery, the patient was symptom-free, and the deformity had been corrected a year later (fig. 2). Preoperative x ray showed the right tibia longer than the left one by 5 millimeters. One year later this difference was reduced to 2 millimeters. The pathological study of the bone specimen did not reveal any structural abnormality.

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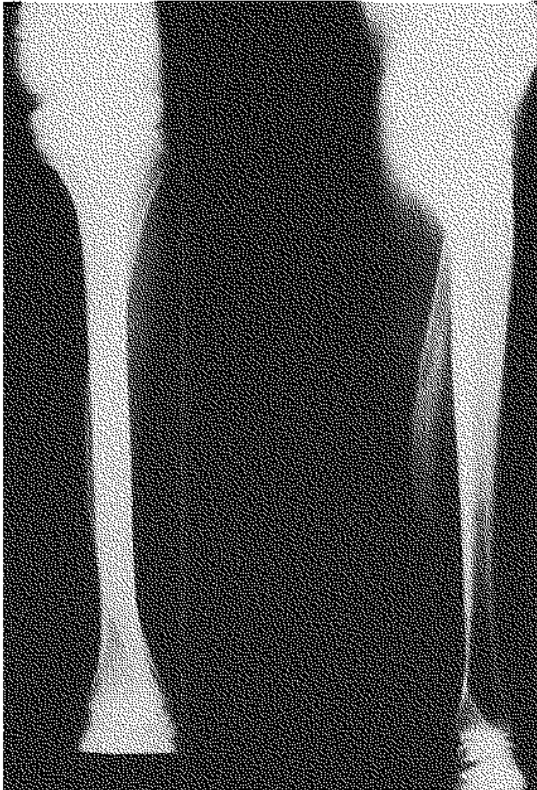


Fig. 1. — Anteroposterior and lateral x ray of the affected leg. Note the curvature of the fibula which is enlarged and elongated.



Fig. 2. — X ray one year after surgery, showing the total correction of the fibular deformity. The patient is now asymptomatic.

DISCUSSION

Congenital and unilateral angulation of the tibia and fibula have been described in a large number of previous publications (1, 3, 4, 5, 7). Three types of deformity have been reported: two affecting the tibia and fibula, with anterior (1) or posterior (3) bowing, and a third type with isolated congenital posterior bowing of the distal fibula (5). This isolated fibular deformity is benign, regressing spontaneously during early childhood, and is not painful.

The differential diagnosis included neurofibromatosis (4), fibrous dysplasia (7), Exner's syndrome (2) and GH-dependent fibular overgrowth (6).

The case described here could correspond to a

fourth type of deformity not yet described in the literature. Our patient had a progressive isolated deformity of the fibula that appeared at a later stage of growth. The main abnormality was not located at the distal end, but the bowing showed a posterior concavity. The deformity became progressively more severe, compressing the adjacent soft tissues and making the ankle joint unstable.

This type of deformity may require surgical correction in the earliest stages of growth, in order to avoid irreversible damage to the ankle joint. In the present case, a shortening osteotomy improved the deformity, and the patient was symptom-free one year after surgery.

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SAMENVATTING

J. C. ABRIL, A. DÍAZ, T. EPELDEGUI. Fibulaire pachydysostose. Een atypisch geval.

Pachydysostose van de fibula is een geïsoleerde misvorming van het been, gekenmerkt door verlenging en incurvatie. Een publikatie, in de literatuur, beschrijft

4 gevallen, die spontaan gunstig evolueerden tijdens de kinderjaren, zodanig dat geen specifieke behandeling aangewezen was.

De auteurs beschrijven het geval van een twaalfjarige jongen, met deze misvorming, waarbij de angulatie geleidelijk toenam, met secundaire misvorming van de enkel. Een osteotomie was nodig om de assen van de fibula te corrigeren. Eén jaar na de ingreep was patiënt klachtenvrij.

RÉSUMÉ

J. C. ABRIL, A. DÍAZ, T. EPELDEGUI. Pachydysostose du péroné. Un cas atypique.

La pachydysostose du péroné est une malformation isolée, caractérisée par une incurvation et un allongement de l'os. Dans la littérature, un article décrit 4 cas qui régressèrent spontanément pendant la petite enfance; aucun traitement ne fut prescrit.

Les auteurs rapportent le cas d'un garçon de 12 ans, présentant cette déformation, avec une angulation augmentant progressivement, provoquant une déformation de la cheville. Le péroné ne put être réaligné qu'après une ostéotomie. La normalisation fonctionnelle fut acquise un an après l'intervention.