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CONGENITAL SHORT FEMUR

By

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In 1936, *Peabody* had collected from the literature 102 cases in which the femurs were congenitally of different lengths. Thus this deformity was comparatively rare until 1958, since when the frequency has clearly increased along with other deformities of the extremities caused by thalidomide. The problem of the cause of a congenital hypoplastic, deformed, or partly missing femur, its prognosis and possibilities for treatment are still largely unsolved. Of course, the limp resulting from the difference in length of the extremities can be effectively treated with an orthopaedic shoe so that in childhood the defect is not very disturbing. According to *Gylding* (1948), various deformities of the femur cannot be clearly distinguished from congenital coxa vara.

The series of deformities of the long bones of the extremities collected from the Helsinki Children's Hospital (*Sulamaa & Ryöppy* 1963) shows that the frequency of such deformities has increased many times since 1957. There was a total of 12 cases treated for difference in length of the femur. These slight deformities were not included in our earlier published series. Eight of these 12 cases were born after 1957.

The aim of the present study of these 12 cases has been to investigate the growth capacity of a congenitally short femur and the effect of treatment on its growth.

MATERIAL

The patients were admitted during the period 1952–1963 and, with the exception of two cases, were under one year old.

On the basis of the shape of the femur our cases can be divided into three main groups:

1. Cases in which the shape of the short femur was more or less normal. There were 6 such cases of *hypoplasia*.
2. Cases in which the diaphysis of the short femur of almost normal shape was bent. There were 3 such cases of *hypoplasia and varus*.

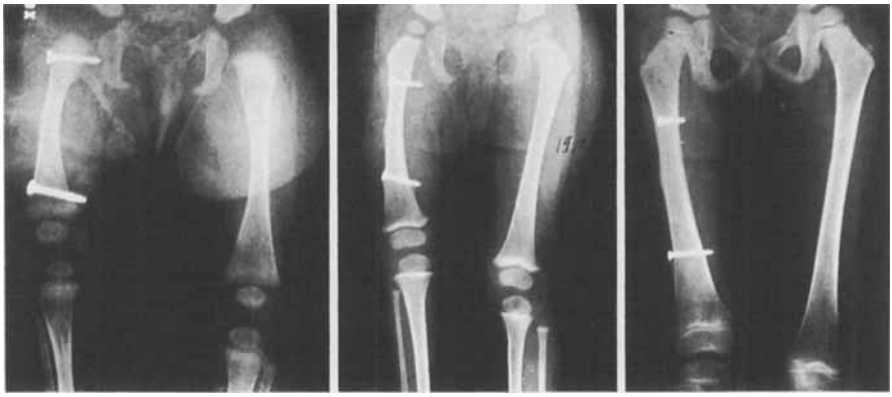

Fig. 1.

Case 2. Radiograms taken at the age of 4 mo, 14 mo, and 4 yrs. The right femur is of normal configuration but slightly shorter than the left femur, which is normal.

3. *Partial defect* or "pseudoarthrosis" of the upper end of the femur, 3 cases.

One of the patients of the first group had other typical "thalidomide deformities" as well, *i.c.* defect of the ulna. In all patients of the hypoplasia group, at the initial stage of treatment, the periosteum was detached and steel screws introduced into the bone close to the epiphyseal line on both the lower end of the femur and the upper end of the tibia.

In cases of the second group the varus position was corrected by osteotomy, and in four cases, including all cases of group three, bone transplantations of some type were carried out in addition.

With the aid of radiograms taken at regular intervals at follow-up the growth of the femur was followed, the femur of the intact side acting as control. The pictures were taken at a distance of 1 metre and the length was measured on the films. Although the method of measurement is not exact, it still does provide, when the healthy femur is radiographed in the same way, a clearly more reliable result than measurement of the extremity itself. The period of observation ranges from one to six years. The difference in length is expressed both in cm and as the proportional length of the shorter femur relative to the healthy one. Table 1 gives information on our cases.

DISCUSSION

The majority of the cases, 6, were referred to the hypoplasia group. In case 6 there were other typical "thalidomide deformities" as well. Since there is, in the present series, a case in which there was aplasia of ulna on one side, it seems evident that the hypoplasia of the femur and the other simultaneous bone defect has a common cause. The cases in which the upper end of the femur is partly missing or deformed (cases 10, 11 and 12) are very similar to the cases of thalidomide de-

TABLE 1

Case	Born	Age of patient at beginning and end of treatment		Operations and age when performed		Difference in length at beginning		at end		Type of deformation
						cm	%	cm	%	
1	1961	10 mo	22 mo	Screw	10 mo	3.3	80	3.3	84	Hypoplasia
2	1959	4 mo	45 mo	Screw	4 mo	2.0	84	3.7	87	Hypoplasia
				Screw	31 mo					
3	1955	57 mo	79 mo	Screw	57 mo	1.6	94	2.5	92	Hypoplasia
4	1958	17 mo	65 mo	Screw	20 mo	2.3	88	3.8	88	Hypoplasia
				Screw	32 mo					
5	1957	2 mo	25 mo	Screw	21 mo	2.2	78	5.4	88	Hypoplasia
6	1960	8 mo	36 mo	Screw	8 mo	2.5	80	3.8	82	Hypoplasia and defect of ulna
7	1959	1 mo	60 mo	Osteotomy	1 mo	2.2	77	5.6	81	Hypoplasia and varus
				Screw	9 mo					
				Screw	18 mo					
8	1958	6 mo	69 mo	Osteotomy	12 mo	4.8	65	4.5	84	Hypoplasia and varus
9	1953	1 mo	75 mo	Screw	12 mo	3.2	62	8.0	70	Hypoplasia and varus
10	1961	6 mo	36 mo	Bonegraft	6 mo	8.0	30	12.7	34	Partial defect and defect of fibula and radius
				Bonegraft	12 mo					
11	1956	5 mo	75 mo	Bonegraft	5 mo	8.7	39	10.7	57	Partial defect and "pseudoarthrosis"
				Screw	40 mo					
12	1958	2 mo	67 mo	Bonegraft	4 mo	6.5	35	17.8	42	Partial defect and "pseudoarthrosis"
				Osteotomy	17 mo					
				Bonegraft	35 mo					

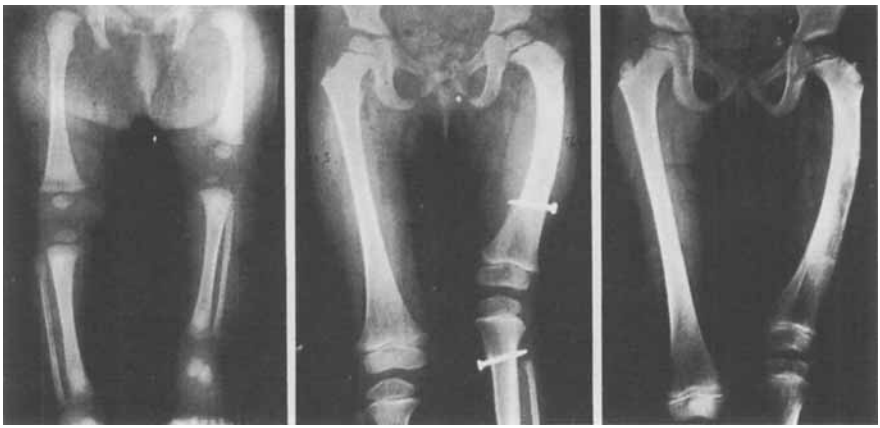

Fig. 2.
Case 7. Radiograms taken at the age of 1 mo, 2 yrs 3 mo and 4 yrs 8 mo. In the neck of the femur there is a slight bend and, moreover, the whole femur is bent.

fects in our previous series. In these cases the difference in length was, as a rule, considerable. Some of the cases of less deformed bent femurs (cases 7 and 8) also resemble in type many deformities caused by thalidomide in other bones. Although a thalidomide aetiology is not with certainty demonstrable in any of the present cases, the time of occurrence of the majority of these cases argues in favour of such a possibility.

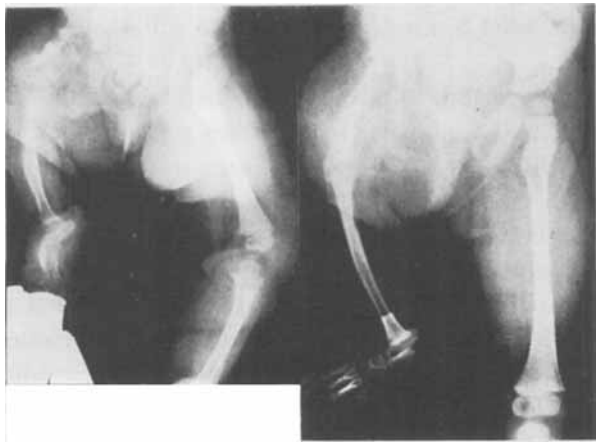

Fig. 3.
Case 10. Radiograms taken at the age of 8 mo and 2 yrs. As a result of two bone graft operations the defective right femur has grown to some extent.

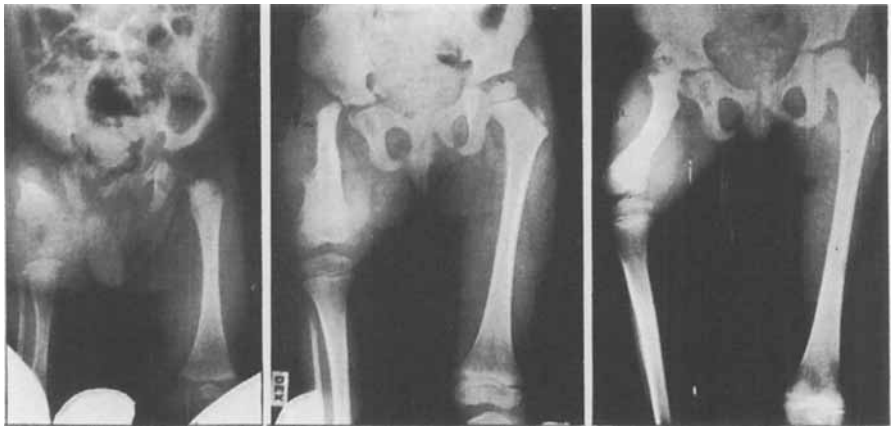

Fig. 4.
Case 12. Radiograms taken at the age of 2 mo, 2 yrs 10 mo and 5 yrs 7 mo. The markedly defective right femur has grown fairly well, thanks to two bone graft operations and one osteotomy.

The shortening of the femur as measured on the radiogram is best illustrated when the length is reckoned as a percentage of the length of the healthy femur. At the beginning of treatment it varied from 80 to 94 in the hypoplasia group. The relative lengths of the varus-shaped femurs ranged between 62 and 77 per cent. A defect of the upper end of the femur (cases 10, 11 and 12), caused a marked shortening. In all cases the shortening was already over 2 cm in infants, with the exception of case 3, in which it was less. The largest difference in length, 8,7 cm in an infant aged 5 months, was due to a defect in the upper end of the femur.

The follow-up period, counted from the beginning of treatment, ranged from 12 to 74 months. In all cases growth led to an increase in the absolute difference in length as compared with the contralateral femur, although the relative difference either remained as before or changed for the better. Detachment of the periosteum and introduction of steel screws does not seem to have stimulated growth to any significant degree. Neither does this small interference seem to have had any negative effect. *Nordentoft & Guldhammar* (1964) came to the same conclusion. A clearly favourable effect on the difference in length was obtained with operative straightening of the varus-shaped femur. A favourable effect on the difference in length by transplantation of bone or epiphyseal cartilage was clearly demonstrable in one case only (case 11). In Peabody's cases the difference in length increased as the pa-

tients grew older. His patients were not operated on. He does not state the difference in length in per cent.

As compared with the results obtained by Pease in sequelae of polio, the results obtained with separation of the periosteum and screws were unimpressive in our cases. When the difference in length was not marked, the slight limp was easily corrected with an orthopaedic shoe, so that the disability did not as a rule cause difficulties of adaptation. At the final stage of growth, the difference in length may also be reduced by stapling or shortening of the longer limb.

Not even a considerable difference in length, such as in our cases 10, 11 and 12, is so disabling as to be an indication for amputation and a prosthesis, as suggested by Blount. These patients have learnt to walk well with the aid of a Thomas' splint and moreover a prosthesis can be attached to the short limb just as well as to an amputation stump. During the few first years of growth an epiphyseal transplant capable of growth may perhaps improve the situation, as shown by Ryöppy in his animal experiments.

SUMMARY

Twelve cases of congenital unilateral hypoplasia of the femur in children are reported. All the patients came for treatment to the Children's Clinic in Helsinki at an early age. The material consists of 6 cases of hypoplasia, 3 cases of "congenital coxa vara" and 3 cases of partial defect of the upper end of the femur. In some cases improvement was obtained by osteotomy and bone graft-operations. The difference in length usually increases in cm but remains the same in percentage compared with the other side. In our opinion there is no indication to perform amputation in early childhood in these cases.

RESUME

Douze cas d'hypoplasie unilatérale congénitale du fémur chez les enfants décrits. Tous les malades ont été en traitement en bas âge à la Clinique de Pédiatrie d'Helsinki. Le matériel d'observation comporte 6 cas d'hypoplasie, 3 cas de coxa vara congénital et 3 cas de défectuosité partielle dans la partie supérieure du fémur. Dans certains cas, une amélioration a été obtenue par ostéotomie et opération de greffe osseuse. La différence de longueur augmente généralement en cm mais

reste proportionnellement la même par rapport à l'autre côté. A notre avis, il n'y a aucune indication pour pratiquer des amputations en bas âge dans ces cas.

ZUSAMMENFASSUNG

Zwölf Fälle von einseitiger Hypoplasie des Femurs von Kindern werden berichtet. Alle Patienten kamen zur Behandling an die Kinderklinik in Helsinki in einem frühen Alter. Das Material besteht aus 6 Fällen von Hypoplasie, 3 Fällen von „kongenitaler coxa vara" und 3 Fällen von teilweisem Defekt des oberen Femurendes. In einigen Fällen wurde eine Besserung durch Osteotomie und Knochenüberpflanzung erreicht. Die Längendifferenz nimmt in cm gemessen gewöhnlich zu, verbleibt jedoch perzentuell die gleiche im Verhältnis zur anderen Seite. Wir haben die Auffassung, dass keine Anzeige zur Amputation im frühen Kindesalter besteht.

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