

## THE PROGNOSIS IN CONGENITAL LOWER LIMB HYPERTROPHY

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Twenty-eight patients with congenital total hypertrophy and ten patients with lower limb hypertrophy with congenital vascular abnormalities are reviewed. The pattern of increase in leg length discrepancy during growth and its influence on surgical management is discussed and the clinical features of the affected limbs are described. In congenital total hypertrophy the maximal increase in leg length discrepancy occurs before the age of 10 years. Those patients who have a leg length discrepancy of more than 2.5 cm at age 4 years are likely to develop significant limb overgrowth that will require eventual surgical correction. In patients with congenital vascular abnormalities the change of leg length discrepancy was variable in degree and unpredictable in pattern, even in those with similar venous anomalies. The outcome for the limb was determined by the nature of the vascular anomaly which should be accurately defined by angiography.

*Key words:* angiomatosis; hypertrophy; leg length inequality

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There are two main clinical problems in the assessment of children with lower limb hypertrophy. The first is to predict at an early age the expected leg length discrepancy at skeletal maturity and the second is to give a prognosis for function and cosmetic appearance of the limb. Although the general features of patients with congenital total hypertrophy (Ward & Lerner 1947, Schwartzman et al. 1942, Bryan et al. 1958) and those with hypertrophy associated with vascular abnormalities (Goidanich & Campanacci 1962, Lindenauer 1971) have been well reviewed, little attention has been paid to the prognostic factors that govern the outcome for the affected limb. Since one of these factors may be the rate of increase of leg length discrepancy during growth we have studied patients with lower limb hypertrophy particularly from this point of view.

### PATIENTS

Two groups of patients with generalised lower limb hypertrophy and leg length discrepancy were studied.

#### *Congenital total hypertrophy*

Twenty-eight patients were studied and classified according to Ward & Lerner (1947).

(a) *Segmental* – 17 patients

Involvement of the whole lower limb without involvement of other structures on that side of the body (Figure 1).

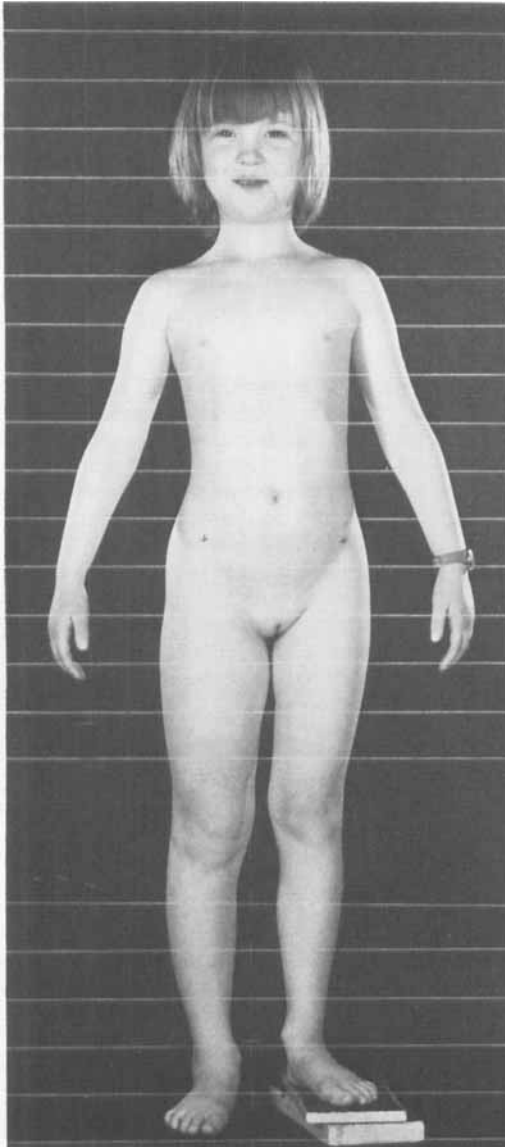
(b) *Crossed* – 1 patient

Overgrowth of one half of the body and segmental hypertrophy of the other side.

(c) *Hemihypertrophy* – 10 patients

Overgrowth of the entire one half of the body (Figure 2).

In these groups there was hypertrophy of all tissues of the involved limb. There is often difficulty in deciding whether one limb or side of the body is hypertrophied or the other atrophied.



*Figure 1. Segmental hypertrophy – right leg. There is no asymmetry of arms or face.*

In this study, patients with neurological deficit on the atrophied side have been defined as hemiatrophy (Penfield & Robertson 1943) and therefore excluded whilst only patients with lengthening of both femur and tibia have been included. Ten patients have been followed from age 4 years to skeletal maturity and the other 18 patients have been followed for varying periods throughout growth.



*Figure 2. Left hemihypertrophy with enlargement of left arm, leg, and side of face.*

#### *Lower limb hypertrophy with congenital vascular abnormality*

Ten patients have been classified according to Lindenauer (1971).

##### *(a) Klippel-Trénaunay syndrome (1900)*

This constitutes the triad of congenital limb hypertrophy associated with varicose veins and cutaneous haemangiomas. Six patients have been studied.

##### *(b) Congenital diffuse arteriovenous fistulae*

Four patients have been studied.

Patients with vascular abnormalities without generalised lower limb hypertrophy were excluded.

## METHODS

Annual radiological and clinical measurements of leg length were made. Photographic assessment of the hemihypertrophy and leg length discrepancy was also undertaken annually (Figures 1 and 2). In hemihypertrophy, bone age showed no significant difference from the chronological age (Dean MacEwen & Case 1967). Full angiographic assessment was made of those patients with congenital vascular abnormalities.

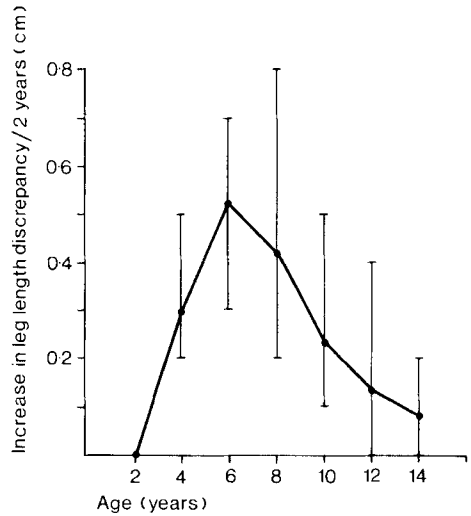
RESULTS

*Congenital total hypertrophy*

*General quality of the limb.* Hypertrophy produced no functional disability in our patients and all had muscles of normal power. Nine patients had cutaneous haemangiomas and one exhibited gigantism of the toes.

*Leg length discrepancy.* The average discrepancy for those patients who did not undergo surgical correction was at 4 years, 1.6 cm (range 0.3 to 2.4 cm), at 10 years 2.3 cm (range 0.9 to 3.3 cm) and at the end of growth 2.6 cm (range 1.1 to 3.7 cm). For six non-operated patients with congenital total hypertrophy who have been assessed regularly from the age of 2 to 14 years, the maximal increase in leg length discrepancy occurred before the age of 10 years. If the average increase in discrepancy per 2 years of growth is plotted against age, the resultant growth velocity curve (Figure 3) confirms the initial rapid increase in discrepancy. In this, admittedly small, group of patients the mean leg length discrepancy at 4 years, 10 years and 14 years (Figure 3) is very similar to that of all patients with the diagnosis of congenital total hypertrophy. Five of the twenty-eight patients underwent procedures to correct leg length discrepancy, and epiphysiodesis is contemplated for a further patient who has leg overgrowth of 4.9 cm at the age of 4 years. All of these patients except one had a discrepancy of more than 2.5 cm (average 2.9 cm) at 4 years of age. The most severe discrepancy was 7.6 cm in a boy aged 11 years who had a lower femoral and proximal tibial epiphysiodesis with an eventual discrepancy of 1.9 cm. The femoral arteriogram in this patient was normal.

*Bone structure abnormalities.* The overall width of both femur and tibia was greater as measured on the X-ray at the points (a) distal femoral epiphysis (b) proximal tibial epiphysis and (c) at the mid-point of each



Mean growth velocity curve: 6 patients with congenital total hypertrophy

Mean total leg length discrepancy:

at age	4 years	1.73 cm	(range 1.2 - 2.2)
	10	2.72	(1.8 - 3.3)
	14	2.98	(1.9 - 3.7)

Figure 3. The mean growth velocity curve is shown for six patients with congenital total hypertrophy regularly assessed from 2 to 14 years of age. The range of increase in leg length discrepancy per 2 years is shown for this group of patients for each age point. The mean total leg length discrepancy is also shown for this group of patients at 4 years, 10 years and 14 years of age.

bone. No other structural abnormality of the bones was noted.

*Lower limb hypertrophy with congenital vascular abnormality*

The clinical features of these patients are summarised in Tables 1 and 2.

*General quality of the limb.* Venographic assessment enabled the patients with Klippel-Trénaunay syndrome to be classified into two categories (Lea Thomas & Andress 1971). In the first category, five patients (A - E) exhibited superficial varicosities associated with normal or dilated valveless deep veins

Table 1. Clinical features of 6 patients with Klippel-Trénaunay syndrome

Patients	Age years	Leg length discrepancy cm			Arteriogram	Venogram
		Tib	Fem	Total		
A	6	+ 1.6	+ 0.6	+ 2.2	No	Dilated valveless sciatic vein Superficial femoral vein normal
	8	+ 1.2	+ 0.7	+ 1.9		
	9	+ 1.2	+ 0.3	+ 1.5		
B	26	+ 2.6	+ 0.5	+ 2.9	No	Dilated valveless deep and superficial veins
C	17	Bilateral		+ 0.5	Normal	Normal deep veins Dilated superficial veins
		R	L			
D	8	+ 2.4	+ 3.1	+ 5.4	Normal	Normal deep veins Dilated superficial veins
E	7	- 0.1	- 0.1	- 0.2	Sciatic	Dilated valveless deep veins
	15	- 0.3	- 0.3	- 0.6	artery	Dilated superficial veins
F	2	0	+ 0.8	+ 0.8	No	Diffuse cavernous angiomata
	4	0	+ 2.4	+ 2.4		
	8	- 0.1	+ 1.9	+ 1.8		

- = Affected limb shorter

+ = Affected limb longer

Table 2. Clinical features of 4 patients with diffuse congenital arteriovenous fistulae

Patient	Age years	Clinical	Arteriogram	Leg length discrepancy cm	Progress
1	16	Limb hypertrophy A-V bruit	Diffuse A-V fistula	1.9	Below-knee amputation for ulceration and pain
2	20	Limb hypertrophy A-V bruit Branham's test + ve Pulsating veins	No	2.2	Above-knee amputation for leg ulcer haemorrhage
3	24	Limb hypertrophy A-V bruit Pulsating veins	Diffuse A-V fistula	3.7	Recurrent ulceration of foot
4	19	Limb hypertrophy Varicose veins Increased foot pulses	Normal	4	No foot ischaemia or ulceration

whereas in the second category, patient F had a limb extensively involved with cavernous angiomata (Figure 4). In the former, the function of the limb was good although the superficial varicosities resulted in problems of venous stasis and were cosmetically undesirable (Figure 5). The cutaneous angiomata (port-wine stain) produced significant cosmetic disability in two of our patients. One patient had gigantism of the toes. In contrast, patient F (Figure 6) had a poor quality limb, massively hypertrophied, with knee flexion and ankle equinus contractures. She subsequently developed disseminated intravascular coagulation secondary to clotting within her extensive venous malformation.

Three patients with diffuse arteriovenous



Figure 4. Venogram of patient (F) showing diffuse involvement of lower leg with cavernous angiomata.

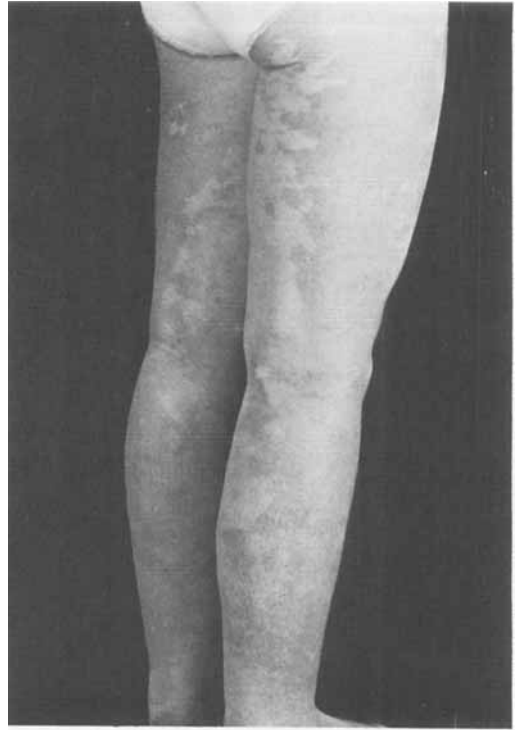
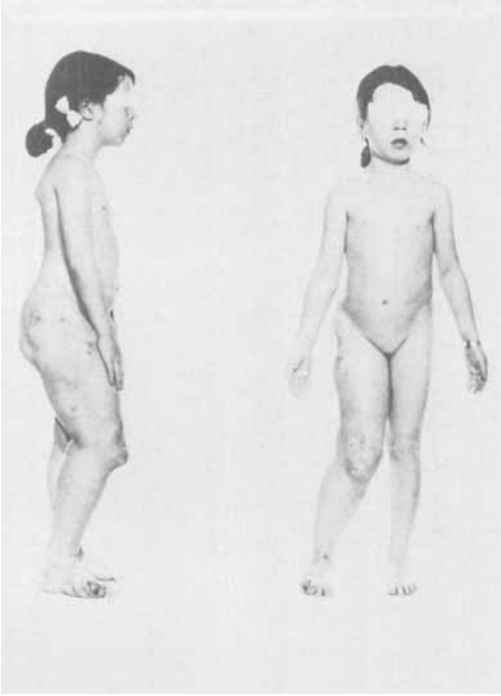


Figure 5. Bilateral Klippel-Trénaunay syndrome (patient C). Normal deep veins on venogram. Good quality limbs although with extensive cutaneous haemangiomata.

fistulae developed distal ischaemic ulceration for which two required amputation. Patient 4 was spared this complication and a clinical diagnosis of multiple hypo-active fistulae was made in the presence of a normal arteriogram.

*Leg length discrepancy.* In patients with Klippel-Trénaunay syndrome there was a wide variation in leg length discrepancy even in patients with similar venous anomalies. The maximum discrepancy was 5.4 cm at the age of 8 years (patient D). Patients A and F exhibited a decreasing discrepancy with growth.

*Bone structural abnormalities.* The bones were not thicker and in patient F the affected bones were thinner. In this patient, the increase in length was all due to femoral



*Figure 6 Massive hypertrophy of right leg associated with diffuse intra-muscular cavernous angiomata.*

overgrowth and irregularity of both lower femoral and upper tibial epiphyses was noted radiographically.

## DISCUSSION

Dean MacEwen & Case (1967) in a review of 32 patients with congenital hemihypertrophy, 26 of whom had leg length discrepancy, stated that 65 per cent of their patients have had or will require epiphysiodesis. Figure 3 shows that in six patients followed from 2 to 14 years the rate of increase in discrepancy slows markedly after 10 years of age. This indicates that the eventual discrepancy at the end of growth was less than would have been predicted at the age of 10 years. We would thus delay operative correction of leg length discrepancy until skeletal maturity and then consider surgery only for those patients

with a discrepancy of more than 3.8 cm, thus avoiding unnecessary epiphysiodesis. We would favour subtrochanteric femoral shortening at the end of growth which allows accurate correction of discrepancy without the development of growth abnormality which not uncommonly follows epiphysiodesis (Hostrup & Pilgaard 1969). Epiphysiodesis should be reserved for those patients who have a major incapacitating discrepancy (i.e. more than 4.5 cm) some years prior to natural epiphyseal closure when femoral shortening would normally be undertaken. The exact timing of epiphysiodesis and the number of epiphyses to be fused should be determined by reference to a growth prediction chart (Anderson et al. 1963). Accurate correction should be possible as our findings suggest that for patients with congenital total hypertrophy there is little increase in discrepancy after the age of 10 years.

In the patients with Klippel-Trénaunay syndrome, a variation in leg length discrepancy was noted, even in those with similar venous anomalies. The rate of increase in discrepancy was unpredictable and two patients exhibited a decreasing discrepancy as growth continued. In two of the patients the limb overgrowth was due to an increase in the length of the femur or the tibia alone.

In those patients with congenital vascular abnormalities, it is the nature of the vascular lesion that governs the outcome for the affected limb. Angiography allows an accurate differentiation to be made between those patients with normal or diffusely dilated deep veins, those with extensive intramuscular cavernous angiomata and those with diffuse arteriovenous fistulae (Lindenauer 1971). We have described the different clinical features associated with these three types of vascular abnormality. In patients with Klippel-Trénaunay syndrome with normal or diffusely dilated deep veins, the limb quality was good and we would not agree that limb overgrowth is a major problem in management (Letts 1977). Varicose veins may successfully be

excised in the presence of competent deep veins. In contrast, the patient with extensive intramuscular cavernous angiomas exhibited gross limb hypertrophy and developed a life-threatening complication. In congenital diffuse arteriovenous fistulae, the outcome for the limb depends almost entirely on the size and activity of the fistulous communications and their amenability to surgical correction (Cotton & Sykes 1969).

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