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The Orthotic Management of the Congenitally Short Lower Limb – A New Appliance

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ABSTRACT

Aim: To describe an appliance used for equalisation of severe congenital lower limb length discrepancy for patients who refuse to undergo any operative correction but wish to walk and look better.

Method: The appliance which is a combination of an ischial bearing brace and a below knee prosthesis was fitted to four patients, aged between 12 and 31 years, with congenital shortening of the lower limb ranging between 17 to 27 centimetres. The diagnosis was proximal femoral focal deficiency in 2 patients, hypoplastic femur in 1 and tibial hemimelia in the fourth patient. The appliance could accommodate existing deformities of the knee, ankle and foot without any operative correction as well as equalise the lower limb length. It is modular in construction and could be easily assembled from off-the-shelf components.

Result: The appliance improves the gait as well as the appearance, could be fitted to the patient without the prior need of operative correction and could be well disguised under any loose fitting garment. Four patients with severe congenital shortening of the leg have used it for over three years and are pleased with it. It requires little maintenance.

Conclusion: The appliance is useful for patients with severe congenital shortening of the lower limb who refuse to undergo operative correction.

Keywords: severe congenital shortening, refuse operation, new appliance

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INTRODUCTION

Patients with rare congenital anomalies such as proximal femoral focal deficiency, tibial or fibular hemimelia present with severe shortening of the lower limb, instability at the hip, knee or ankle and deformities of the foot[1,2,3]. Management of such patients involves extensive surgical procedures to stabilise the joints, correct the deformities and equalise the leg length[4]. At times amputation at the knee or ankle level is followed by fitting of a prosthesis to give a functional limb[5,6]. Some patients due to social beliefs or personal reasons are reluctant to undergo operative procedures and refuse amputation of the foot no matter how deformed but would prefer non-operative measures to look and walk better. A simple appliance, which is a combination of orthosis and prosthesis, was designed to equalise the leg lengths, accommodate the existing deformities of the ankle and foot and improve function without the need for any operation. This appliance has not been previously reported to my knowledge.

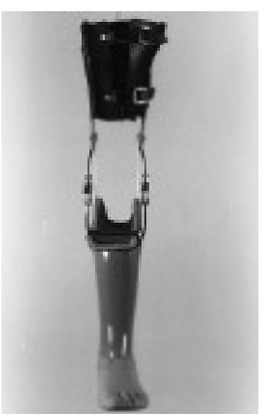

Fig. 1a Frontal view of the appliance.

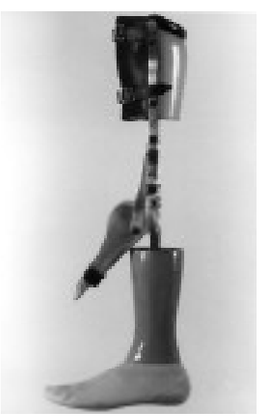

Fig. 1b Side view of the appliance with foot piece in equinus position.

Fig. 1c Side view of a patient with proximal femoral focal deficiency standing with the appliance on.

Fig. 1d Side view of a patient sitting in a chair with the appliance on.

Table I. Details of patients' characteristics.

Case no.	Age (yrs)	Sex	Diagnosis	Hip	Knee (range of flexion)	Ankle	Foot	Associated deformities of the upper limbs	Clinical shorten-ing (cms)	Percent-age shorten-ing
1.	16	F	PFFD (L) Aitken type C	unstable (L)	0^0 to 120^0	10^0 equinus	Absence of a ray leading to a narrow foot	(L) upper limb	25	46
2.	20	F	Hypoplastic femur (L)	stable	0^0 to 125^0	10^0 equinus	no defect	none	27	41
3.	31	F	(L) Tibial hemimelia type II (Jones et al)[14]	stable	0^0 to 90^0	10^0 equinus	Hypoplasia of 1st metatarsal and syndactaly all toes both feet	syndactyly both hands	17	56
4.	12	M	PFFD (R) Aitken type C	unstable (R)	0^0 to 120^0	20^0 equinus	Hypoplastic metatarsal	forearm bones of (R) upper limb	24	50

* PFFD = Proximal femoral focal deficiency

THE APPLIANCE

It is a combination of an orthosis and a prosthesis (Fig. 1a and b), the upper part is an ischial bearing brace (Fig. 2A) with a knee hinge that is joined to the lower part which is a below knee prosthesis (Fig. 2D). A hip joint with a pelvic band, could be added optionally to the proximal part for patients with instability at the hip. The knee hinge (Fig. 2C) allows flexion up to 90^0 to facilitate sitting on a chair (Fig. 1d). The length of the metal uprights (Fig. 2B) of the ischial bearing brace can be varied to compensate for the shortening and allow to place the knee hinge at the same level as the opposite knee.

Two metal struts (Fig. 2F), one each is attached to the inner and outer metal uprights (Fig. 2B) of the ischial bearing brace. These struts support the deformed limb from the knee downwards and have the foot rest attached at its lower end. The top end of the metal struts is attached to the main metal uprights of the brace at a level corresponding to the level of the patient's knee. The struts can swing freely to allow for motion at the patient's knee and to accomodate the flexion deformity. To the lower end of the struts is attached the foot rest (Fig. 2G), which is made from mouldable thermoplastic that could be shaped to accomodate any deformity of the foot. The joint between the plastic foot rest and the lower end of the strut acts like the ankle and can accommodate equinus deformity up to 60^0, further 10^0 to 15^0 of equinus can be accommodated by champfering the front edge of the prosthesis block.

The lower part of the appliance i.e. the prosthesis (Fig. 2D), is made of lightweight material and commercially available Sach foot (Fig. 2E), over which patients can wear the footwear of their choice.

PATIENTS

Four patients, with severe congenital shortening of the leg, have been fitted with the appliance since June 1996. The details of patients' characteristics are given in Table I. There were 3 females and 1 male, their age ranged between 12 to 31 years.The shortening ranged between 17 and 27 centimetres and the percentage shortening varied between 41 to 56 percent. Case 3, with tibial hemimelia type II had the least shortening of 17 centimetres but it constituted 56% shortening of the tibia.

The hip was unstable in cases 1 and 4, both with Aitken type C proximal femoral focal deficiency, but was stable in other two patients. There was some degree of ligamentous laxity of the knee in all patients. Cases 1, 2 and 4 had range of flexion from 0 to 125^0. In case 3, even though the fibula had migrated proximal to the femoral condyles, the knee could flex up to 90^0.

All patients had mild tightness of tendo achilles leading to equinus deformity between 10^0 and 20^0. The foot deformity varied between complete absence of a ray to minor hypoplasia of a metatarsal, resulting in a narrow foot, or just syndactyly.

Three patients case 1, 3 and 4 also had congenital anomalies of the upper limb, they had adapted well to their deformities and all could write. The youngest patient, case 4, was going to school and the other three patients were gainfully employed.

RESULTS

The assessment was purely subjective. All four patients were pleased with gain in height, improved appearance and ability to walk the same distance in less time with

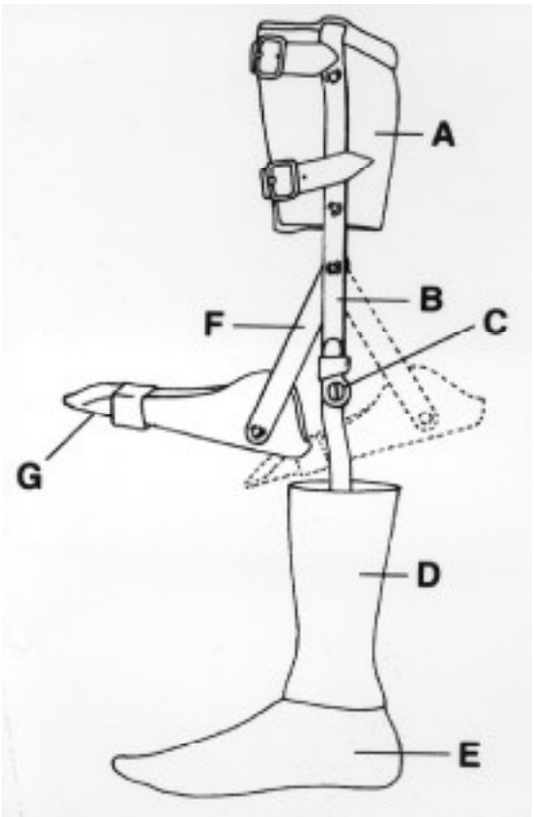

Fig. 2 Line drawing of the appliance showing its various components.

A - the ischial bearing brace.
B - main iron of the tuber bearing brace.
C - the knee hinge.
D - the prosthetic component.
E - the Sach foot
F - the iron strut.
G - the polypropylene foot piece.

the appliance on. They could ascend and descend stairs and sit on a chair after unlocking the knee hinge joint. There were no pressure sores at the heel or around the knee. In three years very infrequent and minor repairs of the appliance were required.

DISCUSSION

Various methods have been described to equalise lower limb length discrepancy but each method has its limitations. Shoe raise is not aesthetically acceptable when the shortening is greater than 5 centimeters[3], epiphyseal stapling or epiphyseodesis is recommended when the anticipated discrepancy at maturity would be in the range of 5 to 8 centimeters[2,3]. Bone lengthening involves extensive surgery and has high incidence of complication should the discrepancy be more than 12 centimeters[3,5,7]. Congenitally deformed or hypoplastic femur or tibia following diaphyseal lengthening of more than 15% of the initial length, behave unpredictably regarding future growth pattern[8,9], and may still leave the patient with unequal leg length at maturity. Moreover lengthening of bone is best suited for patients with stable hip, knee, ankle and a functional plantigrade foot[2,10,11], clinical features which are rarely found in patients with severe congenital shortening of the lower limb.

Several authors have mentioned the use of custom-built orthosis following amputation[2-5,10-12], others advocate preservation of the foot and ankle, especially when in equinus, to get some additional length[5] before fitting custom built extension prosthesis. However when the deformities are severe and fitting of the prosthesis is difficult then amputation of the foot or disarticulation of the ankle is recommended[2,3,5,13,14]. Amputation may not be acceptable to some patients, irrespective of the severity of the deformity or poor function. No appliance has been previously described which could be fitted without prior amputation or corrective surgery.

The appliance, not only helps to equalise the leg length but as an additional benefit provides support to stabilise the hip and the knee which are frequently unstable in congenital deficiencies of the leg. The appliance could be well disguised under any loose fitting garment. Modular construction makes it easy to assemble it from readily available components in any reasonably equipped orthotic or prosthetic department. The foot piece, made from polypropylene, could be moulded to fit the deformity of the foot.

The age of the youngest patient fitted with this appliance was 12 years; it could also be prescribed for even younger children with components made of modern light-weight composite materials, though it would require frequent adjustments in a growing child.

The assessment of the patients was subjective, as their prime requirement was to avoid surgery and look better. Due to lack of facilities, the difference in the rate of oxygen consumption on level walking with and without the appliance was not measured to assess the efficacy of the appliance.

CONCLUSION

The appliance is useful for patients with severe congenital leg length discrepancy who are reluctant to undergo any operative correction of the accompanying deformities of the ankle and foot. Four patients have used it for over three years and are pleased with it. The appliance is modular in construction and could be easily assembled from off-the-shelf components. It requires minimal maintenance.

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