Acta orthop. scand. *53*, 547–552, 1982

PNEUMATIC TROUSER SPLINTS IN THE TREATMENT OF SEVERE OSTEOGENESIS IMPERFECTA

G. MOREL & G. R. HOUGHTON*

Institut Calot, Berck Plage, France

Twelve patients with severe osteogenesis imperfecta have been managed by closed osteotomy to correct lower limb long bone deformity, followed by support in pneumatic trouser splints. The apparatus was well tolerated, fractures were rare, and there was a significant improvement in patient mobility. This conservative method compares favourably with the published series of multiple osteotomies and intramedullary rodding.

Key words: bone diseases, developmental; fracture fixation, internal; orthotic devices; osteogenesis imperfecta; osteotomy; splints

Accepted 17.i.82

Multiple fractures and bony deformity are the most important clinical manifestations of osteogenesis imperfecta (McKusick 1972). Long bone deformity is a characteristic of severe disease (Bauze et al. 1975), and when present in the lower limbs standing and walking is impaired so that disuse muscle atrophy and osteoporosis become pronounced. Abnormal mechanical stresses are borne by the deformed porotic bone which is easily fractured, and may become further deformed (Smars 1961, Falvo et al. 1974, Campanacci et al. 1975).

One of the main objectives of treatment is the early correction of long bone deformity in order to allow the child to stand erect and walk, before secondary atrophic changes become established.

In severe forms of osteogenesis imperfecta, the bones are too fragile to withstand the forces necessary for maintenance of the erect position without support. Support may be provided internally by surgical implants or externally using orthotic devices.

Multiple osteotomy and rodding has gained wide popularity but the method has a high complication rate. In view of these complications a more conservative approach has been introduced (Morel 1971). The deformity is first corrected by multiple closed osteotomies and traction. After the osteotomies have healed, the limbs are supported in light-weight air-filled trouser splints. The child is then stood up in the splints and mobilized. This report is a review of our experience with this form of treatment.

PATIENTS AND METHODS

Twelve patients with osteogenesis imperfecta who have been treated with inflatable lower limb splints and followed up for at least 2 years are included in this series. All patients had severe disease with long bone deformity (Bauze et al. 1975).

The correction of long bone deformity was achieved by closed osteotomy with skin traction to the affected

* Present address: Nuffield Orthopaedic Centre, Headington, Oxford, OX3 7LD, England.

0001-6470/82/040547-06 $02.50/0

Figure 1. Pneumatic splint with the knee held in extension. The outer flutes are in contact with the ground.

limb until the osteotomies became "sticky" and callus was present radiographically. Establishment and maintenance of the erect posture was then achieved with the aid of pneumatic trouser splints.

Techniques

Osteotomy. Closed osteotomy of the tibia or femur is carried out under X-ray image intensification. The site of maximum curvature of the bone is marked on the skin, and the bone is fractured either between the fingers in small children, or over a wooden fulcrum in larger individuals. Multiple osteotomies are usually required in severe deformities involving the entire diaphysis.

Traction. Skin traction only is used. The patients are light weight and skeletal traction is likely to fail owing to cutting out of traction pins through fragile bones. In children under 2 years of age Bryant's traction is sufficient for maintaining bony alignment of both the tibia and femur (Tachdjian 1972). In older children longitudinal skin traction is used. At approximately 6 weeks after osteotomy, union is present clinically, and abundant callus is usually visible radiographically at the osteotomy site. The patient is fitted with the pneumatic splints at this stage, when the necessary measurements have been made after correction of deformities.

Mobilization. After healing of the osteotomies the patient is stood up whilst wearing the pneumatic splints. Initially the patient is supported by the physiotherapist, and then in a body frame or "pulpit" before he progresses to parallel bars and crutches. With children who have been bedridden for many years, care must be taken not to stand the patients precipitously as there will be poor peripheral neurovascular tone and a tendency to faint, as well as poor control of balance. This is minimized by nursing the child on an inclined board during the final healing of the osteotomies, so that the erect position is gradually attained.

The pneumatic splints (Figure 1). The pneumatic trouser splints are worn over underclothes. They consist of a pair of trousers made of inextensible synthetic material (Polyamide 66). The trousers are fitted with zips and laces and incorporated into them are inflatable rubber flutes. When inflated the flutes add rigidity to the garment, but do not constrict the limbs. The flutes can either be continuous across the knee joint to provide

Figure 2. Pneumatic splint with the knee free of support which permits flexion. The lower ends of the splints are strapped to the shoes.

Table 1. Results of treatment in pneumatic splints

Patient	Before splint Fractures	Before splint Mobility	After splint Fractures	After splint Mobility	Time in splint (Years + months)
1	45	W/chair	1	Stick	6+5
2	5	W/chair	1 (1)*	Crutches	3+0
3	8	Crutches	1 (4)*	Stick	8+6
4	5	W/chair	0	Free	5+4
5	20	Bedridden	1	Standing	2+2
6	40	Bedridden	1	Standing	5+4
7	25	Crutches	(1)*	Crutches	4+6
8	8	W/chair	1	Crutches	2+2
9	20	W/chair	1 (1)*	Free	8+0
10	6	Crutches	(2)*	Free	2+3
11	25	Bedridden	4 (2)*	Crutches	6+9
12	12	Crutches	0	Free	3+5

* ( ) fractures sustained during changing or toileting when apparatus not being worn.

stability (Figure 1), or may be interrupted at the knee joint to allow flexion and a comfortable sitting position (Figure 2). The latter type is more suitable for mature patients with quadriceps control of the knee joint.

The splints themselves support the patient with the outer flutes of the trousers in contact with the ground when the patient is standing (Figure 1). In less severely affected children, the lower end of the splint is strapped to the footwear so that all weight-bearing is through the shoes (Figure 2).

Spinal deformity is usually present in severe osteogenesis imperfecta, but this is not a containdication to fitting of trouser splints which are applied to the lower trunk below the apex of the kyphosis or scoliosis.

The patients are maintained in their orthoses until skeletal maturity, when the bones generally become stronger.At that time the children may be gradually weaned out of the splints by progressive deflation of the supporting flutes over a period of months.

RESULTS

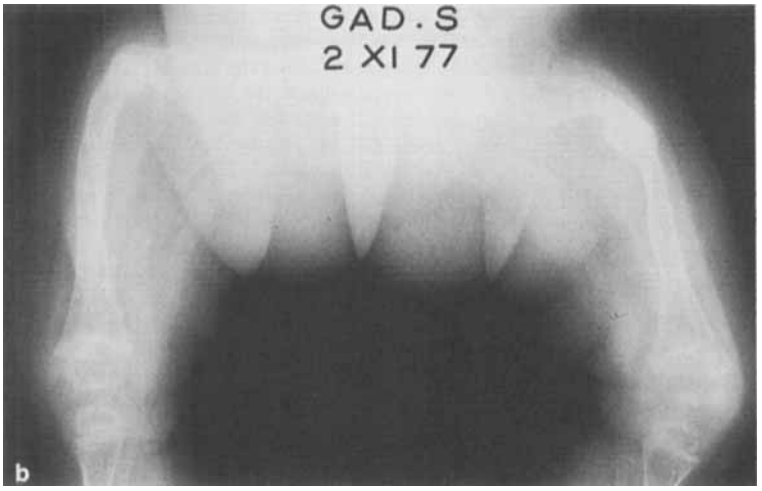
A summary of the results of the conservative treatment regime in 12 patients is shown in Table 1. Before treatment the patients had each sustained between 5 and 45 fractures (mean 20); an average of 2.6 fractures per year. Pneumatic splints were worn by the patients for an average of 4 years and 9 months (range 2 years 2 months to 8 years 6 months). A total of 22 fractures were sustained whilst the 12 patients were being treated in pneumatic splints, an average of 0.33 fractures per year. Half of these fractures occurred as the child was being dressed or washed, when the splints were not being worn. With the exception of Case 7 who was mobile with crutches before and after treatment, the other 11 patients all improved their level of mobility. Four patients were walking freely without aids, whereas before treatment 2 of them were confined to wheelchairs, and 2 required crutches. Two patients walked with a stick, 4 with crutches and 2 who were previously confined to bed were able to stand with support.

CASE REPORT

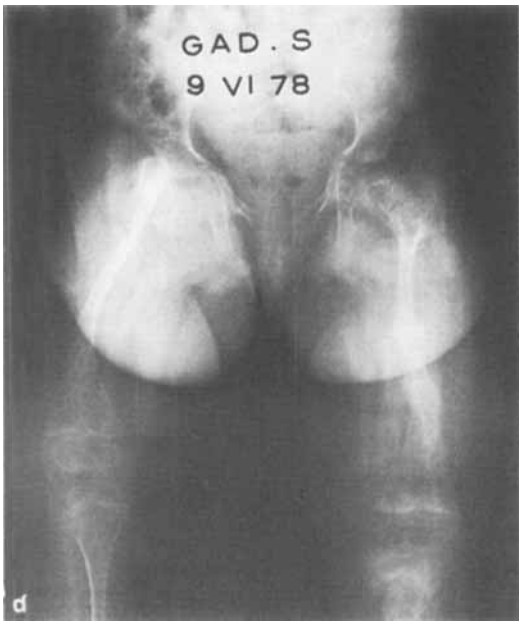
Case 5. A girl aged 4 years and 3 months. Osteogenesis imperfecta was diagnosed at birth when she sustained multiple fractures. During the first 2 years of life she sustained approximately 20 further fractures. When first seen aged 2 years she was irritable, reluctant to move, and could not sit unaided. The skull was flattened from the front backwards, and there were marked long bone deformities (Figure 3a). A radiograph of the femora showed bilateral severe valgus deformity with old healed fractures (Figure 3b). She underwent closed osteotomies of the femora and tibiae and skin traction was applied to both lower limbs for 6 weeks. At this time she was installed in her pneumatic apparatus and stood for the first time aged 2 years and 1 month (Figure 3c). The femora have been held in good alignment in the apparatus (Figure 3d). The patient now spends much of the day out of bed in the vertical position with the aid of the "pulpit" (Figure 3e). She has been in the apparatus for 2 years and 2 months and sustained only

Figures 3a–e Case 5. a) Aged 2 years. Patient confined to her cot with severe long bone deformities and flattened skull and face.

c) Standing for the first time aged 2 years 1 month.



b) Radiograph showing gross varus deformity of the femora, with evidence of healed fractures.



d) Radiograph showing good bony alignment whilst undergoing treatment in pneumatic splints.

e) Aged 4 years. Patient is now able to stand erect with the aid of a "pulpit".

one fracture during that time. There has been a general improvement in her demeanour, and her skull has taken up a more rounded appearance.

DISCUSSION

Multiple osteotomies and intramedullary rodding were first described in 1921 by Springer for the correction of long bone deformity in rickets (Springer 1921). The method was later developed by Sofield and Millar for patients with osteogenesis imperfecta (Sofield & Millar 1959). The method has a high complication rate. There is a high blood loss, with significant mortality (Sofield & Millar 1959, Weber 1977, Marafioti & Westin 1977). Infection is not uncommon (Sofield & Millar 1959, Rebouillat et al. 1969, King & Bobechko 1971, Tiley & Albright 1973, Marafioti & Westin 1977, Moorefield & Miller 1980), and non-union or delayed union may occur (Williams 1965, King & Bobechko 1971, Tiley & Albright 1973, Moorefield & Miller 1980). Problems with rod migration and fractures adjacent to rod ends are very common, requiring revisions (Williams 1965, Rebouillat et al. 1969, King & Bobechko 1971, Rodriguez & Wickstrom 1971, Tiley & Albright 1973, Weber 1977). The introduction of telescopic intramedullary rods has reduced the incidence of revision, but damage to the growth plate with growth arrest as well as damage to the articular cartilage and disassembly of the epiphyseal fixation piece may occur (Rodriguez & Wickstrom 1971, Marafioti & Westin 1977, Finidori et al. 1979, Moorefield & Miller 1980). Although only mentioned specifically as a complication in one series (Moorefield & Miller 1980) extreme porosis invariably occurs around the intramedullary rods.

Pneumatic fluted trouser splints were developed in an attempt to reduce the high incidence of surgical complications (Morel 1971). Standard traditional splints and orthoses are too heavy and cumbersome and are poorly tolerated by children with brittle bones.

There have been no complications directly attributable to the apparatus which is well tolerated by children with bony fragility. Closed osteotomy is relatively atraumatic and blood transfusion is

not needed. Scars in osteogenesis imperfecta are wide and ugly and are avoided if closed osteotomy is performed.

It is our impression that the extreme osteoporosis associated with intramedullary nailing does not occur with external pneumatic support. This is probably because gravitational weight-bearing forces are not eliminated in the splints and osteogenesis is thus promoted in the affected bones. The apparatus enables these severely affected children to stand erect, sometimes for the first time, and this assists their general well being and psychological adjustment to this deforming condition.

ACKNOWLEDGEMENT

We wish to thank Aerazur Constructions, Aeronautiques, 92130, Issy-les-Moulineaux, France, for their help in the development and manufacture of the pneumatic orthoses.

REFERENCES

Bauze, R. J., Smith, R. & Francis, M. J. O. (1975) A new look at osteogenesis imperfecta. A clinical radiological and biochemical study of forty-two patients. *J. Bone Joint Surg.* **57**-B, 2–12.

Campanacci, M., Giunti, A., Leonessa, C., Pagain, P. & Trintain, C. (1975) Pathological fractures in osteopathies and bony dysplasias. *Ital. J. Orthop. Traumatol.*, Suppl. 1, 7–45.

Falvo, K. A., Root, L. & Bullough, P. G. (1974) Osteogenesis imperfecta: Clinical evaluation and management. *J. Bone Joint Surg.* **56**-A, 783–793.

Finidori, G., Padovani, J. P., Rigault, P. & Bensahel, H. (1979) Intramedullary nailing with a telescopic nail in infants suffering from fragilitas ossium. *Orthop. Trans.* **3**, 24.

King, J. D. & Bobechko, W. P. (1971) Osteognenesis imperfecta. An orthopaedic description and surgical review. *J. Bone Joint Surg.* **53**-B, 72–89.

Marafioti, R. L. & Westin, G. W. (1977) Elongating medullary rods in the treatment of osteogenesis imperfecta. *J. Bone Joint Surg.* **59**-A, 467–472.

McKusick, V. A. (1972) *Heritable disorders of connective tissue.* Mosby, St. Louis.

Moorefield, W. G. & Miller, G. R. (1980) Aftermath of osteogenesis imperfecta. Disease in adulthood. *J. Bone Joint Surg.* **62**-A, 113–119.

Morel, G. (1971) Un nouveau type d'appareillage orthopedique: l'appareillage a atteles pneumatiques. *Rev. Chir. Orthop.* **57**, 409–414.

Rebouillat, J., Revelin, P. L. & Brault, J. F. (1969) Treatment of osteogenesis imperfecta with multiple diaphyseal osteotomies. *Pediatrie* **24**, 411–420.

Rodriguez, R. P. & Wickstrom, J. R. (1971) Osteogenesis imperfecta; a preliminary report on resurfacing of long bones with intramedullary fixation by an extensible intramedullary device. *South. Med. J.* **64**, 169–176.

Smars, G. (1961) *Osteogenesis imperfecta in Sweden. Clinical, genetic, epidemiological and socio-medical aspects.* Svenska Bokforlagen Norstedts Stockholm, Scandinavian University Books.

Sofield, H. A. & Millar, E. A. (1959) Fragmentation, realignment and intramedullary rod fixation of deformities of long bones in children. A ten year appraisal. *J. Bone Joint Surg.* **41**-A, 1371–1391.

Springer, H. (1921) Operativer Ausgleich hochgradiger Knocken Verbiegingen durch Zersagung in Scheiben (Segmentierung). Verhandlungen der-Deutschen Orthopadischen Gesellschaft. *Z. Orthop. Chir.* **40**, 118–122.

Tachdjian, M. O. (1972) *Pediatric orthopaedics.* W. B. Saunders Company, Philadelphia.

Tiley, F. & Albright, J. A. (1973) Osteogenesis imperfecta: treatment by multiple osteotomy and intramedullary rod insertion. Report on 13 patients. *J. Bone Joint Surg.* **55**-A, 701–713.

Weber, R. (1977) Complications of the multiple osteotomy technique of Sofield and Millar. In: Symposium on osteogenesis imperfecta, Berck, p. 93–97. Henry, Montreuil-sur-Mer, France.

Williams, P. F. (1965) Fragmentation and rodding in osteogenesis imperfecta. *J. Bone Joint Surg.* **47**-B. 23–31.

Correspondence to: G. R. Houghton, Nuffield Orthopaedic Centre, Headington, Oxford, OX3 7LD, England.