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# Initial Standing of the Osteogenesis Imperfecta Patient

*Steve Baum, C.O.*
*Patricia Hazard, C.O.*

Osteogenesis Imperfecta (O.I.) is more commonly known as "brittle bone disease." The bones are abnormally fragile and will fracture with only minor trauma. Other features of O.I. are weakness, deformity and dwarfism. Bowed long bones and barrel shaped chest are also common.1 Children of this disease have average to above average intelligence and unimpaired hand function.2

The primary focus of this paper concerns a specific case presented to Children's Hospital at Stanford. Rehabilitation Engineering Center. At age two, RH., diagnosed with O.I. type III, its most severe form, was presented to us with a prescription for "a total containment orthosis for support in an upright position." At birth R.H. sustained approximately 22 fractures due to his fragile nature, and great care was required to move or position him. R.H. rests almost exclusively supine and has never borne weight on his lower extremities. He can be held in an upright position, non-weight bearing for only a short period of time before complaining of dizziness. The physiologic advantages of being upright in an erect position include improved kidney and bladder drainage, better cardiopulmonary function, and enhanced bone strength from the effects of gravity on the skeletal system.3,4 Furthermore, an upright posture will enable R.H. to continue toward his developmental goals as a growing child1 and allow him to participate in tabletop activities.

With these goals in mind, the orthotic team met to discuss possible approaches to R.H.'s case The summary of the design criteria is as follows. The orthosis must:

1. be very secure due to the patient's fragile nature;
2. have the ability to be gradually brought up to the fully erect position because patient shows low tolerance for an upright position;
3. have total contact to avoid any uneven loading on the patient's skeletal system: and
4. be portable so it can be taken with the child to school, physical therapy, and home.

Other considerations include ease of Operation for family members as well as school personnel and capability of growth extension since recasting could potentially cause additional skeletal fractures.

It was noted that none of the standing frames that we knew of would be appropriate, because they did not satisfy our criteria. Therefore, we had to create our own design.

## Casting and Model Modification

The casting of the patient was typical though extreme care was exercised to prevent any spontaneous fractures. We made circumferential wraps of the lower extremities and separately cast the patient's torso in a prone attitude through application of plaster splints to his back. Alignment marks for proper adduction and flexion were made on the cast negatives to assist in proper positioning of the orthosis. It should be noted at this time that RH. was cast in his diaper, as this is not standard practice. Since the patient would wear a diaper in the orthosis, its additional width must be accommodated. Because the patient's head control was weak, and the back of his head was flattened due to prolonged supine periods, we also measured for a contoured head support. We filled the negative casts separately for ease of fabrication and transferred the alignment marks to the positive models. The only atypical modifications on the positive models were done to the T.L.S.O. In order to make the orthosis more secure, we slightly straightened and deepened the lateral walls. This also aided the mother in positioning R.H. in the orthosis.

## Fabrication

The orthosis is basically a non-articulating C.T.L.S.H.K.A.F.O. with a baseplate. The description of these components starts at the baseplate and proceeds proximally. The base (Figure 3) was taken from a stock Variety Village Parapodium turned sideways.

Figure 3. Base

Figure 4. Posterior view of telescoping bar and locking pushpins

Attached to the base were two shoe cups with Dacron® and Velcro® fasteners and a small "U" shaped bracket for a telescoping rod. We fabricated the rod from hollow thin walled stainless tubing with a similar but slightly reduced diameter tube to fit inside the outer tube. These were both drilled at 1-1/2" increments and held together with a locking push pin. The rods acted in a fashion similar to an adjustable crutch or cane. The base is a separate unit which attaches to the C.T.L.S.H.K.A.F.O.

The remainder of the standing frame orthosis is the total contact shell. The posterior portions of the K.A.F.O.s and T.L.S.O. are high density polyethylene, 1/8" and 3/16" repectively. The head rest is 1/8" low density polyethylene and the anterior tongues on oth the K.A.F.O.s and T.L.S.O are 1/16" polyethylene. The anterior and posterior components are fastened with Dacron® and Velcro® straps. The headrest was padded with Aliplast® and attached to the T.L.S.O. with a Milwaukee Brace aluminum anterior superstructure upright. In turn, the T.L.S.O. was attached to the K.A.F.O.s with aluminum upright stock. Finally, another small "U" shaped bracket with a locking push pin united the T.L.S.O. to the telescoping rod (Figure 4) .

**Donning and Doffing**

The rationale behind making the orthosis in two parts centered around its relative ease of assembly and attachment to the child in two separate and easily managed steps. The donning procedure begins with laying the C.T.L.S.H.K.A.F.O. section flat with all anterior portions open (Figure 5) .

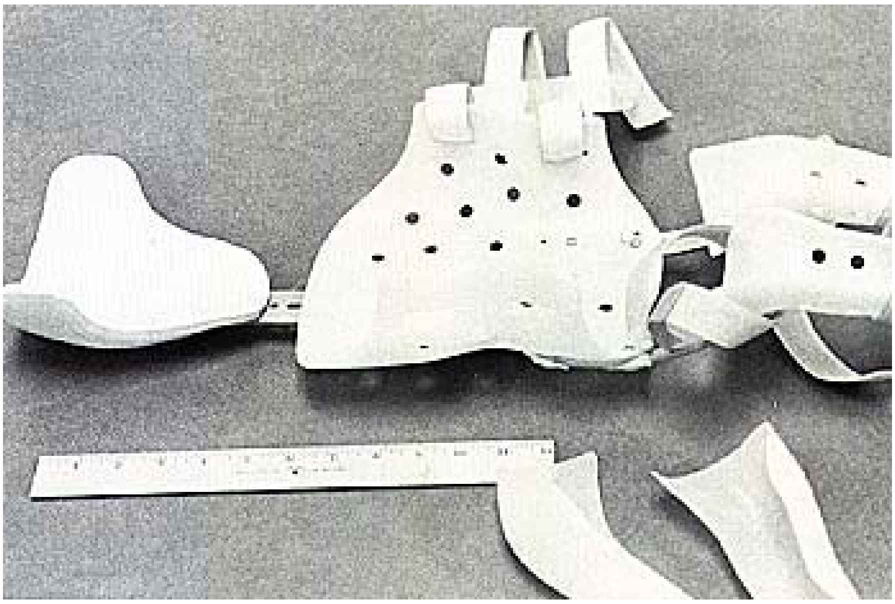

Figure 5. Anterior portions.

We placed the child in the orthosis and secured all anterior shells. We prepared the base with the telescoping bar retracted and the shoe cup straps in their half-closed position. The patient, already secured in the orthosis, is attached to the base by slipping the foot sections into the shoe cups and attaching the single push pin through its bracket (Figure 4) , which unites the T.L.S.O to the telescoping rod. Finally, the shoe cup straps are fully closed.

## Conclusion

Subsequently, the patient has successfully used this orthosis on a daily basis for over three years and has recently needed to be cast for a replacement. The mother is pleased with the orthosis and has made only minor requests for changes to the new orthosis, which includes the need for more air holes to increase ventilation and padding of the entire inner shell. The mother also requested dropping T.L.S.O. walls posteriorly so she can don and doff the orthosis more easily. However, this would make the orthosis less secure, so the anterior T.L.S.O. tongue was made deeper to ensure security.

Many of the particular materials mentioned can be successfully substituted. Many of our choices were made simply by the availability of effective materials. Polypropylene could be used rather than high density polyethylene, or a section of adjustable crutch substituted for the telescoping tube. Obviously, many other options exist.

In summary, this standing orthosis system is a practical and effective means of treating young patients with severe O.I., and can be fabricated with most of the materials that are already on hand.

## Acknowledgements

The design and development of this orthosis was not the work of a single individual. It was the result of a team effort by the staff of the orthotic, prosthetic, and seating/mobility departments of Children's Hospital at Stanford, Rehabilitation Engineering Center.

*Steve Baum, CO., is a staff orthotist and prosthetist at Children's Hospital at Stanford, Rehabilitation Engineering Center, 520 Sand Hill Road, Palo Alto, California.*

*Patricia Hazard, C.O. is the Director of Orthotics & Prosthetics at Shriners Hospital, Lexington, Kentucky.*

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1. Bleck, Eugene E., M.D., "Osteogenesis Imperfecta," Physically Handicapped Children: A Medical Atlas for Teachers, 2nd Edit., Grune & Stratton, Inc., 1982, pp. 4t)5-412.

2. Letts, Merv; Mercer Rang; and Steven Tredwell, "Seating the Disabled," Atlas of Orthotics, 2nd Edit., CV. Mosby Co., 1985, p. 474.

3. Kruger, Leon M., "Children's Orthotics," Atlas of Orthotics, 2nd Edit., CV. Mosby Co., 1985, p. 341.

4. Wolff's Law: "Every change in the form and function of a bone or of their function alone is followed by definite changes in their internal architecture, and equally definite secondary alteration in their external conformation, in accordance with mathematical laws." He believed that the formation of bone results both from the force of muscular tensions and from resultant static stresses of maintaining the body in the erect position, and these forces always intersect at right angles. Excerpt from Rausch and Burke, Kinesiology and Applied Anatomy, 6th Edit., Lea and Febiger, 1978, p. 10.