

CARING FOR AN INFANT WITH KLIPPEL TRENAURY (KT) SYNDROME COMPLICATED BY MICROCYSTIC MALFORMATION OF LEFT LOWER QUADRANT

Brandon's parents brought him to our clinic when he was 3 months old. In the interim since his birth, his parents experienced a wide range of feelings so characteristic of loving parents who have an infant with a congenital disease.

Before beginning therapy we immediately began intense emotional support by giving "lots of hugs", touching both Brandon and his parents, teaching and empowering them. We stressed the fact that Brandon was a beautiful, intelligent, good-looking little boy who happened to have KT and lymphedema. .

Our goals were to teach Brandon's parents to interact with him as a normal boy; teach them how to care for his needs; and teach Brandon, as he grows, how to care for himself.

At 18-20 weeks gestation, a malformation of Brandon's left leg was detected upon routine ultra-sound. Soon after his delivery on January 9, 2001, a thorough medical examination of Brandon indicated he had an extremely large soft tissue over growth of the left foot, especially the dorsum, with massive alteration of the entire leg extending posterior into the buttock with irregularity of the gluteal cleft. In addition, phlebotatic blood vessels were detected within the small vascular stain overlying the left buttock.

Upon measurement, the affected left leg was found to be 2cm longer than the right leg. Further tests concluded the presence of large microcystic lesions containing massive amounts of stagnant lymphatic fluid especially in the foot and lower leg. An MRI performed on February 13, 2001, indicted the extent to which Brandon's left lower quadrant was affected.

Our immediate concerns were the possibility of leakage of lymphatic fluid and the susceptibility of infection creating life-threatening circumstances.

Other critical issues were the potential function of the affected left leg; with 2cm difference in length, would the affected left leg continue to grow more rapidly than the unaffected right leg; KT Syndrome and port wine stain - to what degree was soft tissue and bone affected and how efficiently will fluids move through these affected areas; what was the status of lymphatic connections/anastomoses between the foot and lower leg; how would intensive bandaging affect the integrity of the skin of an infant so small - bandaging had to begin conservatively because of the sensitivity of Brandon's affected foot and very small toes; and lastly, design and availability of gradient compression garments for an infant.

Combined Decongestive Physiotherapy for Brandon began in early April 2001, within days of his first visit. Protocol for manual lymph drainage was designed to move fluids medial to lateral torso and entire leg, anterior and posterior, into left ipsilateral axilla. Skin care consisted of cleansing the entire affected area with Eucerin Gentle Skin Cleanser, rinsing the area with water very well, applying Eucerin Light Lotion followed

by A&D Ointment (emollient), applied over the entire leg and liberally over the foot and between the toes. After applying Stockinette over the entire foot and leg, we then wrapped each individual toe with very thin strips of Aqua-phor impregnated gauze, and Dermagran N Impregnated Gauze over toes or webs that had open or pressure areas.

Thin elastamull strips were then wrapped around each toe to hold protective impregnated gauze in place and to complete adequate compression for the toes. This was very tedious, but it served to prevent Brandon's toes from breaking down during the extended bandaging period. Soft foam "chocolates" or "swell spots" were placed in fibrotic areas such as the calf and dorsum foot. Artiflex "rolls" were placed within Tricofix tubular gauze strips and applied in folds of the ankle and popliteal space. Three (3") or four (4") elastamull rolls wrapped in a spiral fashion from toes to groin held everything in place and protected the little toes. Short stretch bandage rolls were wrapped over elastamull from tip of toes to inguinal, completing compression of the leg for daytime.

Skin care and bandaging of the foot and toes remained the same for nighttime care. A Legacy directional flow garment, full left leg with hip attachment was applied over stockinette. Short stretch bandages were then applied over the Tribute, from toes to waist, to secure its placement and for additional gradient compression.

As reduction progressed we closely monitored Brandon's skin response to bandaging and skin care products in order to ascertain how his skin would react to a future tight fitting daytime gradient compression garment.

It became very clear that skin integrity meant everything to satisfactory garment compliance especially for a fast growing, active little boy. We also felt we needed to be ready for "the right" garment as Brandon increased in mobility and activity, so that muscular movement on his part would increase the flow of lymph out of the calf, foot and buttocks. As Brandon began standing alone, he was placed in a compression class II, with compression beginning on the foot, full left leg with hip attachment and biker short right leg. The crotch was left open for frequent diaper changes. This was perfect for Brandon's transition into crawling and walking, and brought about very good reduction of the entire leg. The foot softened in texture but always remains a challenge compelling us to seek new and different designs for compression and directional flow garments.

As Brandon increased in growth and mobility, the challenge of the garment having been met, shoes and body alignment became our next focus of attention. Children need soft, lightweight, well-made leather shoes for good walking gait and body alignment. After visiting several "shoe makers" it became apparent that these shoes were too heavy for Brandon to pick up his feet while trying to walk.

Brandon's condition and needs called for a "specialty shoe designer" who could "revise Stride Rites" and adapt them to his feet. Arney "the angel" was able to do just that. He revised the left walking shoe to accommodate Brandon's affected left foot and then did a build-up of the right shoe to equalize leg lengths. Brandon liked these shoes and had no problems walking. Arney also revised sandals and sneakers in this manner to accommodate Brandon's growing walking needs.

In September, after several days of 100+ degree weather, Brandon was hospitalized for 5 days with an infection that had begun in the left lower quadrant. Three days of IV antibiotic therapy finally perfused the tissues sufficiently to begin "killing" bacteria to reduce Brandon's body temperature. During Brandon's hospitalization, we began reassessing his entire therapy program by looking at increasing frequency of therapy sessions, revising gradient compression and directional flow garments, decongestion and reduction of lymphedema following infection process by using more creative bandaging techniques, and protecting the skin using more prophylactic skin care products.

Brandon has "bounced back" like any normal 2-year-old boy as if nothing happened. His parents and our clinic remain watchful and more intelligent about ways to care for Brandon's ever growing needs and changes in his life.

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