The controversy over whether sprinter and double-leg amputee Oscar Pistorius should be banned from the 2008 Olympics because of claims his carbon-fiber prosthetics could give him an unfair advantage points to the massive shift in attitudes toward people with limb differences. It wasn’t that long ago that a disability bore the connotation of inferiority or that Dr. Ludwig Guttmann laid the groundwork for the first Paralympic Games. Children and adolescents with limb differences still face a variety of challenges, but with proper medical care and support they can go on to lead active, healthy, and fulfilling lives.

Limb differences occur in about 5 per 10,000 live births, with the most common congenital limb difference being a missing or deformed hand or foot, typically on the left side. The reason for this is unknown. Congenital limb differences are roughly twice as likely to involve the upper versus lower limbs. Upper limb differences frequently are transverse in nature, with congenital absence of all distal segments, while lower limb differences are more likely longitudinal in nature, with the femur, tibia, or fibula being absent or shortened, but the foot still present.

Roughly one-third of congenital limb differences are sporadic; one-third are due to vascular causes including the effects of maternal diabetes, chorionic villous sampling, or uterine abnormalities; and one-third are associated with syndromes such as Holt-Oram, Fanconi, and ectrodactyly-ectodermal dysplasia-clefting. A small number of limb differences are due to teratogen exposure during pregnancy. In a pediatric population, approximately 60% of limb differences are congenital, with the remaining 40% being acquired as a result of disease or trauma. Although infection and cancer play a role in childhood amputations, lawn mower accidents are the most common culprit of amputations in young children.

Limb differences or deficiencies are typically detected in utero or at birth by an obstetrician or pediatrician. Pediatricians seeing an infant with such a diagnosis should consider the possibility of associated deficiencies or abnormalities.

At the 2-year-old Limb Differences Clinic at the Kennedy Krieger Institute, we routinely advise that affected infants be screened for cardiac and renal system abnormalities, as these systems are screening at the same time as the limbs. Genetic counseling also may be helpful in considering underlying syndromes and providing counseling for parents regarding the likelihood of having additional children.

One of the most important decisions for families of children with limb differences is when or whether a prosthesis is needed, and what type of rehabilitation program is needed to optimize function with or without a prosthesis. A multidisciplinary team of professionals from prosthetics, pediatric rehabilitation, orthopedic surgery, and occupational and physical therapy is best to assist in these decisions. If, however, a specialized clinic is not available, the pediatrician may be the first entry point in identifying what services the child needs.

He or she also may have to coordinate care with other providers, track changes or delays in developmental milestones, and provide prescriptions for repairs or revisions to a prosthesis. A general rule of thumb is that a prosthesis is replaced every year until age 5 years, every 2 years until age 12, and then every 3 years.

For children with upper-extremity differences, prosthetic fitting typically occurs when the infant starts sitting up at about 5-6 months. The use of passive prosthesis allows these infants to manipulate objects, which is an important part of their development, and helps to support body weight for creeping and crawling. Early prosthetic use is thought to improve utilization later in life, although some argue that wearing a prosthesis interferes with tactile interaction with the environment.

For children with lower-extremity differences, the first prosthesis is typically fitted at approximately 9 months of age, when the child is ready to pull up to a standing position. Surgical intervention for lower-extremity differences is typically not performed in the newborn period, but at about 1-3 years.

Although addressing mobility and orthotic needs is important, it is essential that the psychological needs of the child and family are met as well. Parents need to accept and deal with the loss of a limb. Ineffective coping may manifest as demanding that a child wear a prosthesis around the clock in an effort to make the problem go away or granting a young child complete autonomy in deciding when to wear the prosthesis, perhaps out of guilt or wanting to make things better. Families do have options regarding prosthetic style; some choose the most lifelike artificial limbs covered in foam and matching skin-tone nylon, while others are comfortable with more durable bare metal limbs.

Although counseling may be needed, providing families with opportunities to meet other individuals facing similar challenges can help these families tremendously in accepting and dealing with limb differences. The Amputee Coalition of America (ACA) offers a service of providing peer visits to interest individuals with limb differences or their family members.

Very young children or siblings may benefit from a line of dolls by Sew Able that have limb differences and accessories such as wheelchairs and parallel bars (www.sew-dolling.com). Having a standard one-liner such as, “I was born this way and can do everything I want,” also can head off lengthy questions from curious classmates.

During the teen years, youngsters may choose to hide their limb difference so as not to be different from their peers, or may refuse to wear their prostheses as a way to exert their independence. However, with the maturation of society regarding persons with disabilities and greater exposure to veterans with missing limbs, recent generations seem more comfortable exposing their limb differences rather than concealing them. One family we’ve known speaks endearingly about their son’s “first red leg.”

The overarching goal of all providers for children with limb differences should be helping these children and their families live active and productive lives. Available technology, combined with the energy of children and increasing community acceptance, provides tremendous opportunities for optimizing function.

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Resources
- Amputee Coalition of America (www.amputee-coalition.org)
- Birth Defects Research for Children Inc. (www.birthdefects.org)
- Helping Hands Foundation (www.helpinghandsgroups.org)
- Limb Differences (www.limbdifferences.org)
- War Amps CHAMP program (www.waramps.ca/champ)

Desonide Foam Improves Atopic Dermatitis–Related Itching

BY KERRI WACHER

PHILADELPHIA — Desonide foam significantly reduced pruritus among pediatric patients with moderate atopic dermatitis compared with the vehicle alone, based on the results of a randomized phase III study of more than 500 children and adolescents.

A multicenter trial included patients at least 3 months old but less than 18 years with moderate AD. Patients were assigned to age-based cohorts: 12 years to less than 18 years, 6 years to less than 12 years, 3 years to less than 6 years, and 3 months to less than 3 years.

In all, 581 patients were included in the study—387 in the desonide group and 194 in the vehicle group. The majority of patients (87%) completed the planned 4-week treatment period—93% of the desonide group and 77% of the vehicle group. There were no differences between the two groups in terms of baseline pruritus scores.

Patients or primary caregivers were instructed to apply the foam once daily (morning and evening) to affected areas over a 4-week period. They were told to include the face and other thin-skinned areas, if affected. Patients and/or caregivers were instructed to assess pruritus for 24 hours prior to each study visit, using a pruritus score table. The investigators did not assess pruritus. Patients who were not able to read and/or understand the score table had their pruritus scored by investigator interview.

In addition, patients or caregivers completed the Dermatology Life Quality Index or the Children’s Dermatology Life Quality Index questionnaire at baseline and again at week 4.

Patients in the desonide group had significantly decreased mean pruritus at week 2 and week 4 compared with those in the vehicle group. No rebound phenomenon was observed 3 weeks after the end of treatment.

The average quality of life measure was significantly improved for the patients on desonide compared with those on vehicle alone at week 4. Dr. Friedlander disclosed that she has received grants for educational activities from Comtechs and several other pharmaceutical companies.