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Valentine Hugs and Kisses are Difficult with Duchenne Muscular Dystrophy

Every Labor Day for the past several years, comedian Jerry Lewis has hosted a telethon to raise money to find a cure for muscular dystrophy. Although most of us have heard of MD, how many of us know that this is not one disease, but a group of at least 20 diseases? The common link in all types of muscular dystrophy is that all are genetic conditions that cause gradual wasting of the muscles, leading to weakness and deformity. In 2001, the Parent Project Muscular Dystrophy launched the first Duchenne MD awareness week to focus attention on one particular form of muscular dystrophy. Five years later, the fifth annual Duchenne Muscular Dystrophy week will be held February 12-18.

The week surrounding Valentine's Day is a time for hugs and kisses and lacey red and pink hearts. The Parent Project points out that 42 muscles are used when hugging someone, and 6 muscles are involved in a simple kiss. By the time children with Duchenne MD have reached their late teens, their muscles have wasted so much that hugging is difficult, if not impossible. And the most important muscle, the heart, is affected by this disease as well.

While all forms of muscular dystrophy are rare, the most common form is Duchenne MD, affecting nearly one boy in every 3000. Although girls can get this disease, it is highly unusual; DMD almost exclusively affects boys, and always results in death by their mid-20s. While most of Duchene's victims die from respiratory failure, anywhere from 9 to 50% die from cardiac failure, according to the Parent Project. There is no cure.

The progression of Duchenne muscular dystrophy occurs in distinct stages. Between the ages of 3 - 7, young boys enter into the early phase, characterized by clumsiness, poor balance, and frequent falling down. Running, climbing stairs, or even getting up from the floor become difficult tasks. As the leg muscles lose elasticity, boys start walking on their toes or the balls of their feet, and appear to be waddling. As they try to maintain their balance, they push their shoulders back and their stomachs out, resulting in a sway-back condition called lordosis.

In the second, or transitional, phase of Duchenne MD, its victims (usually between the ages of 7 and 12 and already diagnosed) have difficulty walking. Sometime during the first two stages, many boys develop enlarged calves. Although the legs appear overly muscular, the increased bulk is due to fat, not muscle cells.

The third phase of DMD occurs between the ages of 8 and 14, and is marked by a loss of ambulation. During this time, a wheelchair is usually introduced for at least part of the time. It is during the teen-age years that most boys suffer the most significant loss of muscle strength, so activities involving the arms, legs, or trunk of the body require some means of mechanical support or assistance. However, fine motor skills usually remain intact, so activities involving the fingers are not affected. Fatigue becomes a serious concern for teen-age boys with Duchenne.

The fourth and final stage, the adult phase, occurs anytime after age 15. While the skeletal muscles were

primarily affected during the third stage, as the young man approaches adulthood, the heart and respiratory muscles typically become compromised.

The risk of heart attack becomes much greater; the young man may experience shortness of breath, swelling of the feet and lower legs, and fluid in the lungs. Either heart or respiratory failure eventually causes death, usually by age 25.

While life is cut short by Duchenne MD, a number of treatments are used to maintain health and mobility for as long as possible. Physical therapy is important in keeping the body flexible, upright, and mobile. Sometimes surgery can help treat the muscle contractures in the legs while the child is still mobile. The use of braces prolongs walking and standing ability, but eventually, wheelchairs become a necessity for mobility and safety. Occupational therapy helps to retrain victims of DMD in conducting activities of daily living, such as dressing oneself or using a computer. Steroids and nutritional supplements may be recommended, although there is little consensus among medical professionals as to the use of these products.

In addition to the wasting away of muscle tissue, DMD affects other parts of the body as well. The spine can be pulled into an abnormal curvature, called scoliosis. Bone density may decrease, resulting in osteoporosis. Muscle cramps sometimes occur. Impaired respiratory function can lead to headaches, nightmares, and difficulty concentrating, as well as a greater risk of infections such as pneumonia. About 1/3 of the boys with Duchenne have learning disabilities, according to the Muscular Dystrophy Association.

Considerable information is available about Duchenne Muscular Dystrophy. The Parent Project Muscular Dystrophy, 800-714-5437 (www.parentprojectmd.org) is one good source, along with the Muscular Dystrophy Association at 800-344-4863 (www.mdausa.org). Additional resources include the Muscular Dystrophy Family Foundation at 800-544-1213 (www.mdff.org), and the Duchenne Muscular Dystrophy Research Fund at 818-692-5500 (www.dmdfund.org). For information about other public health issues, contact the Central Connecticut Health District at 860-721-2824 (www.ccthd.org).