Suited Up For Outdoors

NASA helped design some special suits for special children. There are a number of disorders that make it difficult or impossible for some children to go outside. One such condition is called **hypohidrotic ectodermal dysplasia** (HED). "Hypohidrotic" refers to low levels of perspiration, and "ectodermal dysplasia" means abnormal development of certain tissues derived from embryonic **ectoderm** (teeth, hair, nails, glands). This condition is characterized by a lack of sweat glands. Someone with HED can suffer heat exhaustion, heatstroke, and even death by doing something as simple as going outside to play.

A gene has been identified on chromosome 2 that is involved in some cases of **autosomal** HED. For HED, inheritance is usually X-chromosome linked, but can be autosomal dominant or autosomal recessive.

The hypohidrotic ectodermal dysplasia story dates back to 1875, when Charles Darwin described a peculiar disorder that appeared in each generation of one family's male members. The conditions resulted in poorly developed teeth, sparse hair on the head and body, and excessively dry skin. The individuals observed by Darwin were exclusively males and their symptoms resulted from a mutated gene. This gene was recently identified on the X-chromosome. Females rarely get this form of ectodermal dysplasia because, unlike males who have one X and one Y chromosome, females have two Xs and, therefore, carry a backup gene that can compensate for the mutation. However, there are families where children of both sexes are affected, with no evidence of X-chromosome involvement.

Other characteristics of this disorder include unevenly spaced teeth, no wax in ears, allergies, and asthma. People with this disorder are also highly susceptible to respiratory infections. Skin is often lightly pigmented, thin, and prone to rashes and skin infections. The face may be flat around the nose, the nose itself may be flat, and the eyes may be dry. People with this disorder must stay in a well-regulated environment.

In regards to a well-regulated environment, astronauts face a similar situation when they walk in space. They must protect themselves completely from the harsh environment of space, which can range from 121 ºCelsius [C] (249 ºFahrenheit [F]) to –156 ºC (-250 ºF), depending on the Sun's location. The Sun's ultraviolet (UV) rays can damage skin and eyes, and raise body temperatures to dangerous levels. Protection is essential for astronauts and those with HED. Because of their similar interests in protection from the Sun, NASA and the HED Foundation teamed up to help solve a big problem.

Johnson Space Center's Technology Transfer Office and the HED Foundation created a special UV-protection suit that was developed from space technology. The suit includes a white jacket, pants, gloves, and headgear, including goggles. The external garments protect the patient's sensitive skin from more than 99.9 percent of the Sun's hazardous ultraviolet rays. Underneath the suit, there is a small cooling support system that helps cool the body. Wearing a full-body suit is like wearing a full set of insulation—body heat is trapped, and that raises temperatures. For someone who cannot perspire, that's not good. The cooling unit uses four gel packs in a vest-like garment, and supplies cooling for 2 to 4 hours before needing to be recooled in a refrigerator. This suit allows children with HED to go outside and play—something most children take for granted.

Everyone needs to take care with ultraviolet radiation. Too much exposure, even for healthy people, can result in severe burns and blisters, and increase the risk of skin cancer. For people with HED and related genetic disorders, and for astronauts in space without the filter of our atmosphere, even tiny amounts of UV rays create big problems. Astronauts have worn specially designed space suits for years. This same technology is allowing children with HED to explore their universe in greater detail. Until now, these children had to avoid bright light. They often structured their routines so they were active at night, when less UV danger was present. The special cool suits also allow children with HED to live a more normal life.

In addition to people with hypohidrotic ectodermal dysplasia, the HED Foundation also provides the cool suits to people with **multiple sclerosis**, **spina bifida**, **cerebral palsy**, and other disorders with similar effects.

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