Congenital insensitivity to pain with anhidrosis: a miracle or a curse?

Religion

Have you ever wondered what life would be like without physical pain? Would life be more peaceful? Would we be more aggressive or would we possibly be incapable of enjoying the pleasures of life? We may think we are better off without pain but “ Pain is simply our intrinsic medical adviser to warn us and stimulate us” (H. G. Wells, The Island of Dr. Moreau). Some people have been able to experience this anomaly, the ability to be incapable of experiencing any physical pain. These people are proven examples that pain is in fact necessary. Without pain one an never truly discover their body’s physical limits and may against their knowledge inflict a great deal of pain upon themselves. People with the inability to feel pain have the disease called Congenital Insensitivity to Pain with Anhidrosis (CIPA). CIPA is categorized as a Hereditary Sensory and Autonomic Neuropathic disease. Congenital Insensitivity to pain is the inability to feel uneasy sensations pertaining to an individual from birth. Anhidrosis is the inability to sweat thus causing the incapability to regulate body temperature. HSAN type IV is a genetic disorder caused by multiple DNA mutations.

These mutations occur on the neuropathic tyrosine kinase receptor types one (ntrk1 gene). In normal people the NGR (neuropathic gene receptor) stimulates the growth support and the survival of the autonomic sympathetic neurons as well as nociceptive sensory neurons, which transmit pain sensations to the spinal cord and the brain. Mutations occur in portions of the gene that encode the intracellular and extracellular domain of the protein, which may affect the variability in presentation. Hereditary Sensory and Autonomic Neuropathic (HSAN) type IV causes Congenital

Insensitivity to Pain with Anhidrosis. HSAN IV is the second most common type of HSAN. CIPA symptoms normally manifest at an early age, but can be difficult to diagnose. Infants with CIPA rarely cry from normal ailments, such as not crying when hungry as they cannot sense hunger and they can sleep soundly throughout the night as they cannot sense their needs that need to be met. Parents believe it to be a blessing that their child doesn’t cry until other onset symptoms occur. These symptoms include unexplained fevers, self-mutilation, and developmental delay.

When teething children often chew on anything they can, but since children with CIPA don’t realise the pain of biting themselves, teething becomes dangerous. Many CIPA children begin to bite off their own tongue and fingers when teething; often ripping out their own teeth as well. Around this time, parents begin to realise that these symptoms are not normal for children. People with CIPA can’t live a normal life because of the many unfortunate ailments caused by the disease. Hyperactivity is a common characteristic of children with CIPA, as is eing unable to differentiate between temperatures. Many children die early because it is very easy for them to overheat as they cannot regulate their own body temperature. The lack of sweating causes the skin to become thick and susceptible to skin infections and cellulitis. Their bones deteriorate over a shorter period of time than normal bones. Their joints are overused usually confining patients to a wheelchair (wheelchair bound). An infinite number of patients develop appendicitis and die because they are unable to feel the pain that this would normally cause.

It is impossible to determine the number of CIPA patients, as there is quite a discrepancy in the statistics. Unfortunately, there is no cure for CIPA, and they are very few treatment options. Since it is a rare disorder there hasn’t been a lot of research done on the topic. To regulate the body temperature, some individuals with CIPA have to wear a special cooling vest designed by NASA, and must have a controlled thermostat. These individuals, like 11 year old Kayla Woodhouse, need to have a cooler filled with ice pack re-fills for her vest, and fluids to keep her hydrated.

Other patients with CIPA wear goggles to protect their cornea, which would otherwise become damaged from their habit to scratch themselves. Gabby Gingras is one of the well known CIPA patients who wears goggles to prevent further damage. Gabby also had to have her teeth removed by the age of two to prevent further irreparable damage to her fingers. People with CIPA need constant supervision to avoid future injuries. Life without pain is more difficult than at first glance; it wouldn’t be as wonderful as we would assume. Upon further investigation of CIPA, although it may seem like a wonderful thing t is an extremely dangerous disorder that most often results in premature death. Until we find a cure, people who have CIPA are forced to painlessly suffer. Imagine if humans could suddenly no longer feel physical pain: Pharmaceutical companies, doctors, and manufacturing companies would cry all the way to their banks. Bibliography Axelrod, F. B. , von Simson, G. G. , Oddoux, C. (2008, August 5). Hereditary Sensory and Autonomic Neuropathy IV. GeneReviews. Retrieved November 15, 2008, from, http://www. ncbi. nlm. nih. gov/bookshelf/br. fcgi? book= geneð= hsan4.

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