IS LIFE BETTER WITHOUT PAIN?

(1) Most people agree that pain is very unpleasant, but you may be surprised to know that pain is also useful. Pain functions to do three important things: 1) it causes you to remove yourself from the situation producing the pain, so if you accidently burn your hand on a hot stove, you'll lift your hand; 2) it causes you to protect the damaged body part, so you'll inspect your burned hand, run it under cold water and put a Band-Aid on it if the skin is broken and 3) it teaches you to avoid similar situations in the future so you will be more careful around hot stoves.

(2) Despite the benefits of pain, would life still be better without it? Let's take a look at people who feel no pain and see if this improves their lives. Some people have a rare inherited disorder of the nervous system called CIPA, or congenital insensitivity to pain with anhidrosis. The word *congenital* means that the condition existed from birth and the word *anhidrosis* means that the person's body is incapable of sweating. People with CIPA cannot feel pain because extreme temperatures (hot or cold), tissue damage (cuts to the flesh) and extreme pressure (having your fingers crushed by a slammed car door) are not noticed by the brain as different from other ordinary sensory experiences. They also don't feel the discomfort caused by hunger or a full bladder.

(3) Sensory neurons are responsible for carrying sensations from the body to the brain, including sensations of pain. A gene called NTRK1 is responsible for creating an enzyme that helps sensory neurons form properly during the development of embryos. As well, this enzyme prevents apoptosis, which is a process that causes the sensory neurons to die prematurely. People with CIPA have a mutation of their NTRK1 gene which prevents their sensory neurons from forming.

(4) Nociceptors are the sensory neurons that collect pain information from the body and send it to the brain to be experienced as pain. People with CIPA don't have nociceptors so they can't experience pain. Nociceptors come in various forms: mechanical nociceptors detect excessive pressure to the skin or flesh, thermal nociceptors detect the extremes of hot and cold, chemical nociceptors detect noxious

chemicals and some nociceptors are called sleeping or silent nociceptors because they only send pain signals when tissue damage has occurred. For example, if the temperature is too hot, thermal nociceptor can send signals of pain which are experienced as discomfort, but if the temperature increases to the point that it begins to cause tissue damage, the sleeping or silent nociceptors will activate and begin to add their pain signals to those of the thermal nociceptors. This informs the brain that the danger to the body has increased.

(5) The life expectancy of people with CIPA is lowered and death during childhood is increased. Since they can't feel pain, they often injure themselves repeatedly without knowing they are doing so and they can also let wounds become badly infected because of their lack of awareness. If an infection goes untreated, it can easily lead to the amputation of a limb or if an infection migrates into the blood, the person can suffer a life-threatening condition called sepsis where the entire body becomes inflamed and can go into shock.

(6) Parents of children with CIPA have to take extra care to prevent injury. When babies are teething, they tend to chew on things. Babies with CIPA can easily chew through their tongues or even bite through their fingers because they can't feel pain. Some parents choose to pull out all the baby teeth so that this doesn't happen, though this makes it harder for the child to eat solid food. Parents also have to monitor how much their child eats because they might not eat enough since they experience no hunger pains. It's common for most children to rub their eyes vigorously and

IS LIFE BETTER WITHOUT PAIN?

without a sense of pain or discomfort, children with CIPA easily scratch their corneas and can get serious eye injuries.

(8) Throughout their life, people with CIPA are at risk of fracturing or breaking bones without knowing it. As well, most of us shift around all day as one sustained body position becomes uncomfortable for our joints. People with CIPA stay in the same position for long periods of time which causes a lot of stress on the joints. This eventually leads to joint injuries. Another serious threat comes from overheating. People with CIPA can't tell when they are getting too hot, and when they are, they are unable to sweat to cool off. Excessive core body temperature can lead to seizures and even death if the body isn't cooled in time.

(9) CIPA is a autosomal recessive genetic disorder. This means that the NTRK1 mutation is located on a chromosome that doesn't determine a person's sex. It also means that both parents have to have the mutated gene to give their child CIPA. Though people with only one mutated NTRK1 gene won't get CIPA, they are carriers of the gene and they can pass this mutated copy on to their children. The disease is a rare one because it is unlikely that both parents will have the mutated gene.

Article Questions

1) Describe three reasons why pain is useful.

2) What does CIPA stand for?

3) What is anhidrosis?

4) When the NRTK1 gene is mutated, what is the result?

5) What is a nociceptor?

6) Besides mechanical, thermal and chemical nociceptors, why are sleeping or silent nociceptors important?

7) Describe two things that parents of children with CIPA should be careful to monitor.

8) How does an individual get CIPA and why is the likelihood of this rare?