Inability to Feel Pain

Pain is an integral part of the defense system of the body. It signals that something is wrong, and helps to minimize the physical harm that is done to the body. In the majority of cases, when a person finds something to be painful, they react in such a way to alleviate the pain, resulting in the harm to their body being minimized; however, in certain individuals, the pain is either not felt, or no reaction is observed, resulting in more harm being done to the body [(1)](http://serendip.brynmawr.edu/bb/neuro/neuro06/web1/aschmid.html#1).

Congenital analgesia, also known as congenital indifference to pain, is a rare condition in which there is an absence of pain sensation from birth without the loss of other sensations or demonstrable nerve pathology [(2)](http://serendip.brynmawr.edu/bb/neuro/neuro06/web1/aschmid.html#2). This can result in the individual unintentionally harming him or herself, or in an injury being made worse by the individual not realizing its severity. The first report of the condition was made in 1932, concerning a man who acted as a human pin cushion. Since then, there have been fewer than 100 reported cases in the United States [(3)](http://serendip.brynmawr.edu/bb/neuro/neuro06/web1/aschmid.html#3). The exact cause of the condition is unknown; the nerves appear to be normal and functioning properly in the majority of cases. The condition is believed to be hereditary [(2,](http://serendip.brynmawr.edu/bb/neuro/neuro06/web1/aschmid.html#2), [3)](http://serendip.brynmawr.edu/bb/neuro/neuro06/web1/aschmid.html#3). The higher frequency of occurrence among the children of consanguineous parents suggests that the responsible allele is recessive, although there have been reports of what appears to be an autosomal dominant version of the condition. The reports of the autosomal dominant variation of congenital analgesia are too infrequent to come to any real conclusion at this time, but it is possible that the condition may be caused by more than one allele [(5)](http://serendip.brynmawr.edu/bb/neuro/neuro06/web1/aschmid.html#5). Congenital analgesia is sometimes associated with auditory imperception, along with a trisomy of one or more chromosomes in the 13-15 group; however, this association is by no means proved [(3)](http://serendip.brynmawr.edu/bb/neuro/neuro06/web1/aschmid.html#3).

Another condition with similar effects is congenital insensitivity to pain with anhidrosis (CIPA) [(4)](http://serendip.brynmawr.edu/bb/neuro/neuro06/web1/aschmid.html#4). Unlike congenital analgesia, CIPA appears to be caused by a mutation in the NRTK1 gene, which codes for a nerve growth factor-specific tyrosine kinase receptor, which, as the name suggests, affects the growth of nerves. Individuals with CIPA have a highly decreased number of small myelinated and unmediated nerve fibers, to the point that they may be entirely missing in the epidermis [(4)](http://serendip.brynmawr.edu/bb/neuro/neuro06/web1/aschmid.html#4). These missing fibers may explain why pain is not felt, but do not explain why most other sensations are felt, with the possible exception of temperature. In a large portion of the reported cases of CIPA, the individual lacks sensitivity to temperature changes. This may or may not be related to the anhidrosis portion of the condition, which is the inability to regulate one's body temperature by producing sweat. In individuals with CIPA, the sweat glands appear to be normal, but no sweat is produced when elicited by a variety of stimuli. This may be due at least in part to the individual's inability to sense temperature changes, as sweating is also not seen when the person with CIPA develops a fever, even when it is in excess of 109 degrees Fahrenheit [(4)](http://serendip.brynmawr.edu/bb/neuro/neuro06/web1/aschmid.html#4).

A third condition involving the inability to feel pain is hereditary sensory and autonomic neuropathy type 5 (HSAN5) [(6)](http://serendip.brynmawr.edu/bb/neuro/neuro06/web1/aschmid.html#6). Individuals with HSAN5 have a loss of pain sensation as well as the ability to sense temperature changes, as with individuals with CIPA. Unlike those with CIPA, individuals that have HSAN5 do not have anhidrosis; they responded normally to stimuli that were meant to induce sweating. In individuals with HSAN5, there is a virtual absence of small myelinated afferent fibers, but only a small reduction in the number of small unmyelinated fibers. The nerve conduction velocities of the tested patients were normal, but no response was seen when attempting to record an evoked response over the spine via tibial nerve stimulation. Unlike both CIPA and congenital analgesia, some individuals with HSAN5 only had a loss of pain sensation in their limbs [(3](http://serendip.brynmawr.edu/bb/neuro/neuro06/web1/aschmid.html#3), [6)](http://serendip.brynmawr.edu/bb/neuro/neuro06/web1/aschmid.html#6). There was also evidence in some cases of HSAN5 of degeneration of the unmyelinated axons, suggesting that the condition, unlike CIPA and congenital analgesia, may progress over time. Like congenital analgesia, HSAN5 appears to be caused by an autosomal allele or set of alleles, with the majority of cases appearing in children of parents who are first cousins, or a closer relation [(6)](http://serendip.brynmawr.edu/bb/neuro/neuro06/web1/aschmid.html#6).

With all of the conditions, there is a high incidence of self-mutilation from infancy, as the individuals with the conditions cannot feel the pain that they are causing themselves, and thus do not know when to stop. Injuries to the lips and tongue are relatively common during teething, as the child does not realize that they are harming themselves. As the individuals age, problems from injuries such as broken bones become more common, as they do not realize that the bones are broken, and may attempt, for instance, to walk on a broken leg, feeling nothing more than slight uncomfort. In individuals with CIPA and HSAN5, there is a high incidence of complications from infection, as they cannot detect the pain or that they have a fever, and may not realize that they are ill until the infection has become serious. In the past, there was a high chance of death during childhood for those with any of these conditions, due to the inability of doctors to diagnose and treat the conditions, and the inability of the individual to detect illness at an early stage, along with their inability to realize when they were harming themselves. Today, survival rates have increased, although raising a child with one of the conditions mentioned above is very difficult. While they are young, they require near constant supervision to make sure that they are not harming themselves, and must visit a doctor more frequently than children without one of the conditions. Once they are older, the majority of the supervision may end, but extra care must be taken to prevent them from accidentally injuring themselves [(3](http://serendip.brynmawr.edu/bb/neuro/neuro06/web1/aschmid.html#3), [4](http://serendip.brynmawr.edu/bb/neuro/neuro06/web1/aschmid.html#4), [5](http://serendip.brynmawr.edu/bb/neuro/neuro06/web1/aschmid.html#5), [6)](http://serendip.brynmawr.edu/bb/neuro/neuro06/web1/aschmid.html#6).

At this time, the exact causes of congenital analgesia, CIPA, and HSAN5 are not known, although mutations that tend to occur in individuals with these conditions have been noted. I am surprised that there hasn't been more research done on the subject, as it could lead to advances in other areas, especially if the nerve degeneration in HSAN5 occurs on a similar manner to nerve degeneration in other diseases. Further research into the causes of CIPA and congenital analgesia may also prove fruitful, as it could lead to new methods of blocking pain.