Title: Congenital Insensitivity to Pain with Anhidrosis *GeneReview* – CIPA: Related Challenges and Risks
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CIPA: Related Challenges and Risks

Individuals with CIPA lack NGF-dependent neurons, including NGF-dependent primary afferents and sympathetic postganglionic neurons [Indo 2012]. Pain is an unpleasant sensory and emotional experience. Systemic responses of the sympathetic nervous system often accompany emotional responses. Emotion is a strong feeling characterized by various complex reactions with both mental and physical manifestations closely related to activities of the sympathetic system. The 'fight-or-flight' response illustrates a strong emotional state associated with an extreme excitation of the sympathetic nervous system.

When animals, including humans, are exposed to danger or trauma, stimuli or contexts associated with the danger or trauma become learned triggers that unleash emotional responses [LeDoux 1996; LeDoux 2000]. From birth, normal individuals experience emotions such as fear whenever they are exposed to danger or trauma in daily life. These emotional experiences then induce so-called 'fear conditioning' by pairing the stimuli or contexts with danger or trauma. Therefore, emotional responses serve a protective role by producing aversion to contexts associated with danger. Individuals with CIPA cannot detect various noxious stimuli nor trigger emotional responses to noxious stimuli in the same way as normal individuals because they lack NGF-dependent neurons [Indo 2012]. Accordingly, they may be impaired in their ability to modify their behaviors in order to protect their bodies and maintain homeostasis. Because of this they are always at a disadvantage, with threatened survival.

References

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