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Unusual Skin Symptoms in Autonomic Dysfunction: A Pictorial Essay

This article is a collection of pictures with descriptions showing skin symptoms in children with various types of autonomic dysfunction.

"Slap" patches

These reddened areas of skin occur frequently in children with autonomic dysfunction, and may appear on any part of the body. They are typically extremely hot to the touch but not painful. While they may be more likely to occur during periods of stress, they often pop up unexpectedly for no apparent reason. They are caused by blood rushing to a specific area, filling the capillaries and dilating the blood vessels.

What is interesting about these patches is that they are commonly restricted to one side of the body at any given time, and typically only affect one or two areas of the body at a particular time. When they recur, they may recur on the same side of the body or in the same location, or they may be on the other side of the body and in a completely different location.



Child with Vaccine-Induced Encephalopathy and Autonomic Dysfunction with a "slapped cheek" patch on his face while well. It is common for these patches to only occur on one side. In this child, they usually appear on the right cheek, but occasionally show up on the left.



The same child as above with both the "slap" patch on his cheek and a red hot right arm. He was not feeling well at the time of the picture. The redness typically lasts for a few minutes to several hours, and always clears up overnight.



"Slap" patch or hot, reddened mark on a leg in a child with Cerebral Palsy secondary to Hypoxic Ischemic Encephalopathy, along with the skin rash keratosis pilaris (chicken skin or goose flesh). The keratosis pilaris is very common in children and probably unrelated to this child's Autonomic Dysfunction. This child gets slap patches in a wide variety of locations on her body, with no usual side or location. They usually disappear within an hour.

Red Ear Phenomenon

Oddly enough, many children with autonomic dysfunction have periods when one ear turns bright red and hot to the touch. This flushing does not necessarily affect any other part of the body. These red ears may occur in times of stress, but often appear sporadically for no clear reason. They are similar to the "slap patches" described above, but appearance of these red ears is so common that they warrant their own category.

There is a painful red ear syndrome that has been described in the medical literature, but there have been very few documented cases and no clear cause, other than the idea that nerves are not regulating blood flow to the ear appropriately. Whether red ear syndrome is the same thing as the hot red ears seen in children with autonomic dysfunction is yet to be established.



Red Ear Phenomenon with no additional facial flushing in a boy with Vaccine-Induced Encephalopathy. The phenomenon only affects his right ear.



Red Ear Phenomenon on a girl with Cerebral Palsy secondary to Hypoxic Ischemic Encephalopathy. She routinely has either ear affected, and occasionally both at the same time during periods of stress or autonomic crisis.

Erythematous blotching

Erythematous blotching consists of red hot splotches that occur as a result of dilated blood vessels and capillary congestion. They differ from the above categories in that they tend to be discontinuous, forming a more rash-like blotching instead of a solid red area. They occur most commonly on the arms, legs, lower face, neck, and upper torso.



A child with moderate Autonomic Dysfunction secondary to Hypoxic Ischemic Encephalopathy experiencing an autonomic crisis due to severe pain. This picture shows erythematous blotching of the neck and upper torso. She also experiences a fast heart rate (>180 beats per minute), high blood pressure, and extreme irritability during crises. Once she is given pain medication and oxygen to help stabilize her vitals, the blotching disappears completely, lasting only about five minutes after the crisis has concluded.



Same child, with erythematous blotching on her arm.



Same child, with erythematous blotching on the face. Note that she is screaming and extremely irritable.

Flushing of the Whole Face

Flushing, like the majority of these processes, is caused by blood rushing to the head and congesting the blood vessels. It causes redness and warmth in the entire face. In some cases it may occur in tandem with sweating. Flushing is common during periods of stress on the body, such as illness, pain, or exposure to warm temperatures.



Child with Vaccine-Induced Encephalopathy and Autonomic Dysfunction with flushing during a period of illness.



Flushing and sweating in a baby with an undiagnosed disorder and Autonomic Dysfunction. His temperature during these periods is actually low (about 96F) and his heart rate is elevated. The sweating does not occur when his temperature is normal or high. His symptoms are relieved somewhat by laying down, but can last for hours and occasionally even for days.

Mottling or Cutis Marmorata

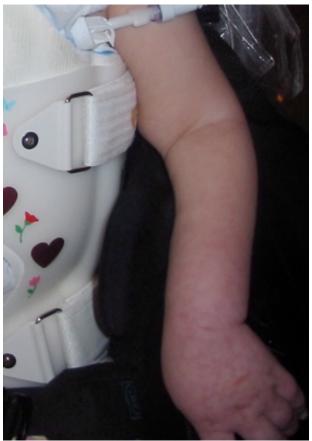
Many children with autonomic dysfunction have mottling of their hands and feet. While this is especially common in children who also have muscle tone problems (such as in cerebral palsy) or Down Syndrome, it can be seen in any child with autonomic dysfunction. In mottling, the nerve supply to the capillaries in the skin is unstable, and some capillaries swell while others contract. There are red "lines" where the blood vessels are dilated, and pale areas where they are contracted, causing the mottled effect.

The mottling tends to get significantly worse in cold weather.

Mottling or Cutis Marmorata is also common in newborn babies due to immaturity of the nervous system and is considered a benign condition. There is a more serious version called Cutis Marmorata Telangiectatica Congenita that tends to occur in tandem with congenital deformities, but this is rare.



Mottling or Cutis Marmorata in a child with Cerebral Palsy secondary to Hypoxic Ischemic Encephalopathy and Autonomic Symptoms.



Another child with Autonomic Dysfunction and Cerebral Palsy secondary to Hypoxic Ischemic Encephalopathy with very mottled hands, extending upwards to mid-forearm.

Purple or Cyanotic Extremities

Many children with autonomic dysfunction, especially those who have problems with muscle tone, also get cold, purple feet and hands due to poor circulation. While this problem resembles Raynaud's Syndrome and generally does worsen in the cold, in children with autonomic dysfunction, it is generally thought to be a part of the autonomic dysfunction and not a separate disorder.

The red flushing, blotches, and patches described above are caused by the dilation of blood vessels. In the case of cold, purple, or cyanotic hands and feet, the blood vessels constrict or spasm and represent reduced blood flow to the area.



Cold, purple feet in a child with Cerebral Palsy secondary to Hypoxic Ischemic Encephalopathy and Autonomic Symptoms.



Cold, purple hand in another child with Autonomic Dysfunction and Cerebral Palsy secondary to Hypoxic Ischemic Encephalopathy. Note the difference in color between the mother's skin on the left and the child's hand.



Same child as above, with cyanotic nails, purple fingers, and white splotches that represent areas of blood vessels that are completely contracted.