



PHACE syndrome among children, its clinical features and nursing management

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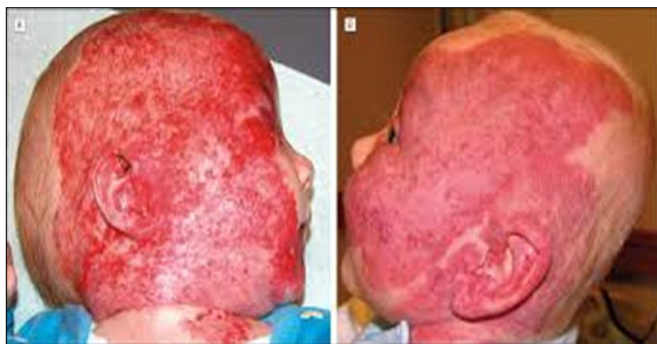
Abstract

PHACE syndrome is an association between infantile hemangiomas and malformation of eyes, heart, major arteries and the brain. The acronym Phace stands for Posterior fossa brain malformations.

Keywords: PHACE Syndrome, hemangiomas

Introduction

Children are an asset for future National abundance. Talking statistically, there are 73.8 million children in the world of aged 0-17 in years and about 30 % of the population is comprised of this age group. As children under five are more vulnerable to various infections, they are at the risk of high mortality and morbidity rates all over the world. Considering, them as an asset to nation, it is the responsibility of the particular government to provide comprehensive health services to all the Children to promote the health and wellness.



PHACE syndrome is the association of a large hemangioma, usually on the face or neck, in combination with one or more other abnormalities. Every infant diagnosed with PHACE syndrome has different medical needs. Some have mild symptoms while other have more severe symptoms.

PHACE syndrome is uncommon but may have been misdiagnosed in the past. It affects girls nine times more often than boys.

Initially this syndrome was described as an association of large cutaneous hemangiomas of the head and anomalies of the cerebral vasculature by Pascual-Castroviejo in 1978. Subsequently term PHACE was coined by Ilona Frieden.

Definition

PHACE is an acronym that refers to a group of abnormal medical findings. When these occur together, doctors can diagnosis PHACE syndrome.

PHACE defined:

- Posterior fossa – brain malformations that are present at birth
- Hemangioma – This usually covers a large area on the skin of the head or neck (greater than 5 cm). The term "segmental" is sometimes used to describe these hemangiomas.
- Arterial lesions – abnormalities of the blood vessels in the neck or head
- Cardiac abnormalities/aortic coarctation – abnormalities of the heart or the blood vessels that are attached to the heart
- Eye abnormalities.

Causes

The exact of the PHACE syndrome is unknown.

There has been a great deal of research to understand the cause of PHACE Syndrome and Research is ongoing to find the cause of all vascular anomalies including PHACE Syndrome.

The abnormalities associated with this syndrome are thought to be due to errors that occur very early during development.

There may be a genetic component involved and studies are underway to investigate this idea.

Signs and symptoms

The most common sign of PHACE is the presence of a large (greater than 5 centimeters wide) hemangioma - a benign vascular tumor - on a child's neck, face or scalp.

For the purposes of developing a diagnosis in a child suspected of having PHACE (see Testing & Diagnosis), those abnormalities and symptoms are divided into two categories.

- a. Major criteria.
- b. Minor criteria.

Symptom of Major criteria

- Aortic arch anomalies such as coarctation of the aorta.
- Anomalies (e.g., narrowing, abnormal growth, absence) in the major arteries of the head and brain (the cerebral arteries).
- Continued presence after birth of the trigeminal artery, an artery in the brain that is present during fetal development

but which typically disappears before birth.

- Anomalies of the cerebellum.
- Aneurysms in the blood vessels of the brain or heart.
- Abnormalities in the structures in the back of the eye (e.g., retina, fovea, optic nerve).
- Abnormalities in the blood vessels of the retina or other parts of the eye.
- Defects in the sternum (breastbone), such as cleft sternum.

Symptoms of Minor criteria

- Ventral septal defects (gaps in the wall between the heart's left and right ventricles, or pumping chambers).
- Continued presence after birth of fetal arteries other than the trigeminal artery.
- Pituitary gland or thyroid anomalies.

Diagnosis

If the medical history and the actual exam of the hemangioma look typical for PHACE Syndrome, more tests are ordered to confirm the diagnosis. These tests may include:

- Ultrasound
- Magnetic resonance imaging (MRI)
- Magnetic resonance angiography of the brain (MRA)
- Echocardiogram
- Eye exam by an eye doctor
- Other tests may be needed for diagnosis and treatment.

Treatment

Treatment of PHACE syndrome is individualized and will depend upon a variety of factors including the organs that are affected. The hemangioma(s) may require medical therapy to control the growth. The child may need other therapies, depending on what other organs or structures are involved.

Management

PHACE syndrome needs to be managed by a multidisciplinary team of experts familiar with the treatment of hemangiomas and special considerations that arise when a child has PHACE. Additional specialties such as cardiology, ophthalmology, neurology, and neurosurgery may need to be involved.

This team of experts needs to follow the infant initially to manage acute issues and complications but it is very important for this same team to follow the patient long term to evaluate development and any long term issues, pay close attention to how these children develop throughout the school age period.

Complications

- Hemangioma is near the eye it may cause problems with vision development in that eye.
- The facial hemangioma on these infants is more likely to break open and bleed.
- Migraine headaches.
- Seizures.
- Delay in development.
- Delayed speech.
- Very rarely ischemic strokes.
- Aortic arch abnormalities.

Role of a Nursing to manage the child with PHACE syndrome and their family

- Nurse play a key role in dealing the patients with PHACE syndrome and even can help the affected families to deal with the Physical and developmental changes of the affected child.
- Nurse should alleviate the anxiety of the parents of affected children by helping them to cope up with the situation by providing support groups and counseling Services.
- Nurse should help the families to collaborate with the health care team for child proper development and treatment.
- The team of Nurses pays close attention to how the children with the PHACE syndrome develop throughout the school age period.
- Nurse must play a key role in creating awareness among the people about the condition.
- Nurse must support the patients and families of those with the condition and conduct the research for the better contribution to the affected society.

References

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