Ask the Doctor

Pamela Pilcher*, MD, discusses the brain and it's affects on movement. This is the first in a series of discussions of the intricacies of the brain and Cri-du-chat syndrome (5p-).

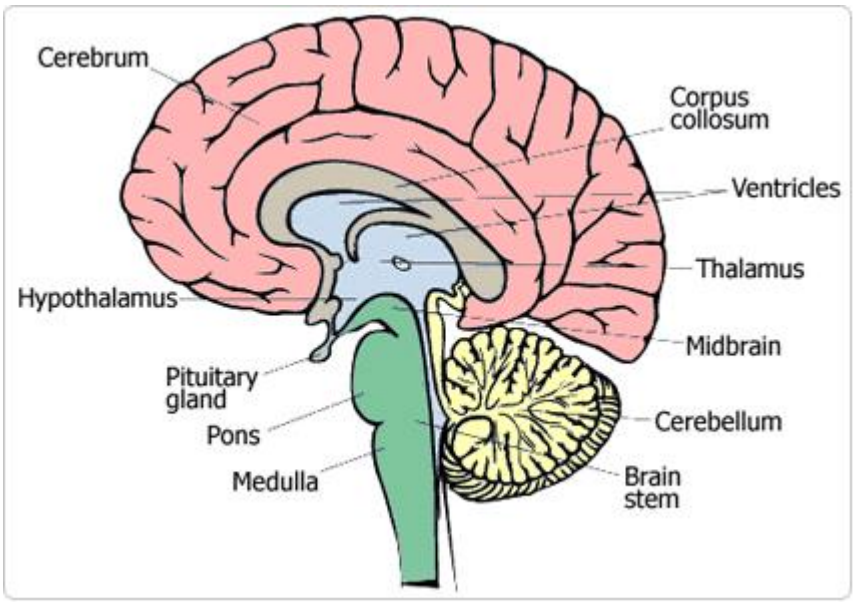
INTRODUCTION: Cri-du-chat is a deletion syndrome. The deletion of genetic material occurs on the 5th chromosome of its short arm (p). The amount of genetic material lost on the 5th chromosome varies in every child making the spectrum of possible disorders large.

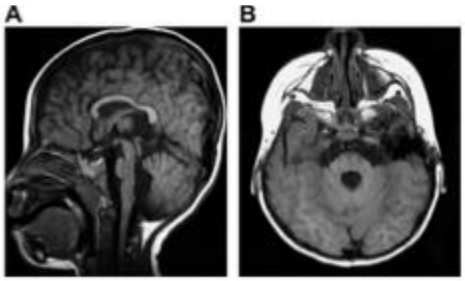
TOPIC: NEUROANATOMY OF CRI-DU-CHAT (5P-) BRAIN AND ITS AFFECTS ON MOVEMENT

Definition: "*Hypoplasia* – is the incomplete development or underdevelopment of a tissue or an organ."

Medical literature review of Neuroanatomy of Cri-du-chat (5p-) brain based on MRI results appears to substantiate changes in the formation of the Pons, Medulla, Corpus Collosum, Cerebellum including its Vermis, and the bony cranial base of the skull which supports these areas. Hypoplasia of the Pons seems to be the most consistent finding and, at times, is accompanied by Cerebellar hypoplasia and Corpus Collosum atrophy. The changes in these areas are a result of incomplete development or underdevelopment of the tissue and have an impact in the development of movement.

Image Comparision of Normal Brain and MRI of 5p- with underdevelopment of the Pons:





MRI "T1-weighted images at the age of 1 year and 6 months showing pontine hypoplasia with no abnormality in other parts of the brainstem or cerebellum." T. Ninchoji, et.al/Brain & Development 32 (2010) 571-573.

Discussion of delayed movement from hypoplasia:

The hypoplasia that has been documented in the medical literature brings to light the areas of the brain that are affecting the movement development in the Cri-du-chat population, thus, allowing us to begin to understand the possible cause for their movement dysfunction. Based upon normal physiology, in a general sense, the Cerebrum, Midbrain, Pons, Medulla, Cerebellum, and Basal Ganglia form a complex system of checks and balances that regulate and define movement of the trunk and extremities. The underdevelopment of the Pons, and Cerebellum in the Cri-du-chat brain appears to disrupt the ability of these structures to modify movement on a minute-to-minute basis causing observable movement disorder. The ability to execute movement with precision or the ability to integrate simple motor tasks into complex motor skills, such as riding a

bicycle, appears to be delayed because of the hypoplasia in these areas. The ability of the Cerebellum to coordinate input from multiple sources and "to compare what you think you are going to do (according to the motor cortex) with what is actually happening down in the limbs (according to proprioceptive feedback), and correct the movement, if there is a problem" is placed at a great disadvantage with its underdevelopment.

(http://www.bioon.com/bioline/neurosci/course/cerebell.html)

CONCLUSION:

The brain is a highly specialized organ that is interdependent upon each of its parts for the completion of any and all movement tasks. When full growth and development of any of its parts is delayed, abnormal movement is observed and perpetuated.

REFERENCE LIST

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6. "Pontine Hypoplasia in 5p- Syndrome: A key MRI finding for a diagnosis." Takeshi Ninchoji, Jun-ichi Takanashi, et al. Department of Pediatrics. Kameda Medical Center.

929 Higashi-cho Kamogawa-shi. Chiba 296-8602, Japan. Received 27 March 2009. Received in revised form 4 July 2009. Accepted 7 July 2009.

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Dr. Pilcher has put together the information below to help you better understand how each area of the brain was made to function. As you read the medical articles she has written, please, refer back to this information for better comprehension.

The BRAIN and its Function

1. **Midbrain**: contains Cranial Nerves (CN) 3 & 4. The Midbrain is associated with the Superior Cerebellar Peduncles. Assists with postural control via Basal Ganglia and Thalamus.
2. **Pons**: contains CN 5, 6, 7, and 8. The Pons is associated with the Middle Cerebellar Penduncles. The Pons is where the fibers from the Cerebral Cortex cross over to the other side of the body and then into the Cerebellum.
3. **Medulla**: contains CN 9, 10,11, and 12. The Medulla is associated with the Inferior Cerebellar Peduncles.
4. **Cerebellum**: Coordination center for all muscle movement of the body. The hemispheres control purposeful limb movements. The output of the Cerebellum to the Cerebral cortex is excitatory. Its input is balanced by the Basal Ganglia. Its pathways effect the *same side* of the body.

   Vermis of Cerebellum - controls posture of the trunk of the body.

5. **Basal Ganglia** – a collection of nuclei that modify movement in association with the Cerebellum and Thalamus. The output of the Basal Ganglia to the Cerebral cortex is inhibitory. Its input is balanced by the Cerebellum.

6. **Corpus Collosum:** a band of nerve fibers joining the brain hemispheres.

7. **Cerebrum** includes the following lobes: Frontal, Temporal, Parietal, Occipital Lobe. Its pathways affect *the opposite side* of the body.

8. Each lobe has specific functions and are described as follows:
9. Frontal Lobe:

a. Social Judgment: inhibition of behavior.
b. **Motor: planning and execution of motion.**
c. Eye Movements
d. Language Production: Comprehension is intact. Speech is coherent.
e. Executive Function: Concentration, orientation, abstraction, judgment, mood, and inhibition of primitive reflexes.

B. Temporal Lobe:

a. Hearing.
b. Auditory pathways.
c. Memory
d. Emotion.
e. Language comprehension

C. Parietal Lobe:

a. Sensation
b. Spatial relationships
c. Ability to read and write.
d. Ability to make arithmetic calculations.
e. Vision
f. Visual and Cognitive Attention.

D. Occipital Lobe:

a. Visual pathways.
b. Visual recognition.

9. **Cranial Nerves (overview of main function):**

CN 1 - Olfactory Nerve (Function: Smell)

CN 2 - Optic Nerve (Function: Sight)

CN 3 - Oculomotor Nerve (Function: Controls the muscles involved in eye movement except the Superior Oblique and the Lateral Rectus.)

CN 4 – Trochlear Nerve (Function: Moves Eye Down and Out**.** If the nerve is injured or the muscle is weak, the eye when looking towards the nose will ascend upward towards the eyebrow instead of downward. When reading or looking down, the eyes will see

double images. The head will be tilted away from the eye that is ascending when it looks toward the nose.)

CN 5 - Trigeminal Nerve (Function: *Sensation* – receives sensory input from the face to the brain. *Movement:* controls the adduction and lateral motion of the jaw through the Masseter, Temporalis, and Medial and Lateral Pterygoid.)

CN 6 – Lateral Rectus (Function: Moves Eye Out to the Side)

CN 7 - Facial Nerve (Function: *Sensation* - Carries information from the anterior 2/3rds of the tongue and parasympathetic fibers of the head and neck ganglia. *Movement:* Moves facial muscles, Stapes (ear bone), Hyoid bone. Produces tears and saliva.)

CN 8 - Vestibulocochlear Nerve (Function: Hearing and Equilibrium**)**

CN 9 - Glossopharyngeal Nerve (Function: *Sensation* – receives information about blood pressure from the Carotid Sinus and sends this information to the Medulla which when stimulated decreases arterial blood pressure and heart rate. Sensory input is also received from Pharynx, Tonsils, Middle Ear, and Posterior Tongue. *Movement:* elevates the Pharynx (e.g. back of the throat).

CN 10 – Vagus Nerve (Function**:** *Sensation* – relays information from body organs in abdomen back to the brain. *Movement:* controls posterior tongue and soft palate, heart rate, breathing, voice reasonance, and small and large intestine contraction.)

CN 11 - Spinal Accessory Nerve (Function: *Movement* – controls the Trapezius and Sternocleidomastoid muscle. The Trapezius muscle elevates the shoulder girdle and moves the scapula towards the spinal column. The Sternocleidomastoid muscle elevates the shoulder girdle and acts in lateral flexion of the head to the shoulder on the same side.)

CN 12 – Hypoglossal Nerve (Function: *Movement* -controls tongue muscles. It is responsible for

speech and the act of swallowing.)

REFERENCE:

Twelve Cranial Nerves: The 12 Pairs of Nerves That Emerge Directly From the Brain. https://12cranialnerves.wordpress.com