

INTRODUCTION:

Cri-du-chat (**5p-**) 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.

TITLE: AIRWAY FROM NOSE TO LUNGS

Pathway of Air from Nose to Lungs:

Air enters the nose and sinuses, then, moves into the throat where it flows through the upright epiglottis down into the trachea and on into the lungs.

(<http://www.healthhype.com/wp-content/uploads/respiratory.png>)

Common Abnormalities of the Airway from Nose to Lungs in (5p-) Population:

Air enters the nose and sinuses and moves into the throat. As the air moves to the larynx – the air meets with resistance from the narrowed passageway of soft tissue with low tone. As the remaining air comes to the floppy, underdeveloped epiglottis, which is unable to be fully upright, only some of the air is able to pass into the trachea and then into the lungs. Thus, the 5p- child may not get the full breath of air he inhaled into his lungs, and he may tend to be chronically hypoxic.

DESCRIPTION OF CRI-DU-CHAT AIRWAY from Anesthesiologists:

In the article entitled: “Airway Evaluation by CT Imaging for Cri-Du-Chat Syndrome”¹, the anatomy of the Cri-Du-Chat Syndrome is described: “In general, patients with cri-du-chat syndrome exhibit a range of laryngeal abnormalities, including hypoplastic (*e.g. underdeveloped*) larynx, narrow laryngomalacia (*e.g. narrow larynx caused by soft tissue*), and diamond-shaped larynx, as well as epiglottal abnormalities, including long, curved, floppy, hypo- plastic epiglottis and hypotonic epiglottis, all of which present risks of difficulty in airway management during anesthesia.”

ANATOMY and FUNCTION:

- 1) **LARYNX**- “a tube shaped organ in the neck that contains the vocal cords.”
(www.medicinenet.com)
- 2) **Image of Larynx:**
https://www.cedars-sinai.edu/Patients/Programs-and-Services/Head-and-Neck-Cancer-Center/Treatment/Images/larynx_model_web-80443.jpg

- 3) **Hypoplasia:** “underdevelopment or incomplete development of a tissue or organ. It refers to an inadequate or below-normal number of cells” (<https://en.wikipedia.org/wiki/Hypoplasia>)
- 4) **Malacia:** “abnormal softening of tissue” (<https://en.wikipedia.org/Malacia>)
- 5) Vocal Cords (visualized):
http://www.entmags.org/archive/images/Vocal_Cord_Paralysis.jpg
- 6) **Functions Associated with Larynx:** “Its primary function is to protect the lower airway by closing abruptly upon mechanical stimulation, thereby halting respiration and preventing the entry of foreign matter into the airway. Other functions of the larynx include the production of sound (phonation), coughing, the Valsalva maneuver, and control of ventilation, and acting as a sensory organ.” (Laryngeal Anatomy. Author: Rishi Vashishta, MD; Chief Editor: Thomas R. Gest, PhD...updated August 31, 2015. Medscape.
- 7) **Image of Epiglottis:**
<http://www.intechopen.com/source/html/41621/media/image1.jpeg>
- 8) **Function of the Epiglottis:** “The epiglottis is a flexible flap at the superior end of the larynx in the throat. It acts as a switch between the larynx and the esophagus to permit air to enter the airway to the lungs and food to pass into the gastrointestinal tract.” (www.innerbody.com/image_digeov/dige02-new2.html)
- 9) **Hypotonia:** low muscle tone which may also involve loss of strength.

CONCLUSION:

In Cri-Du-Chat Syndrome, varying abnormalities of the airway are unique and may be common in this population. The Anesthesiologists that authored this article made this recommendation based upon these abnormalities: “We strongly recommend that the preoperative evaluation of the airway by CT imaging is of great utility for patients with Cri-Du-Chat Syndrome” as X-rays may not visualize these abnormalities with the same clarity. (see End note below)

¹ “Airway Evaluation by CT Imaging for Cri-Du-Chat Syndrome” by H. Arisaka, S. Sakuraba, M. Matsumoto, H. Kitahama, M. Furuya, K. Yoshida, and J. Takeda.