



A Review of Craniosynostosis in Communication Disorders Practice and its Effect on my Family

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Abstract

Craniosynostosis is a congenital malformation of the sutures of the skull which fuse too early in childhood which does not allow the skull to expand appropriately. The cognitive and physical consequences of this fusion can last a lifetime. There are several types of craniosynostosis depending on what suture fuses and these have a variety of long-term consequences. The diagnosis is typically made after birth and some of the sequela can be appended if surgery is completed in a quick manner. The children born with craniosynostosis will have long-term complications of which the author, Ms. Peters, is readily aware given her family connection. Working with someone with this disorder can be challenging, but rewarding.

Keywords: Hearing; Maxiofacial; Genetic

Introduction

Definition and Cause

The skull is made up of five bones: frontal, parietal, temporal, occipital, and sphenoid. These bones fuse together to create a suture (where two bones meet). This fusion begins at approximately two years of age and completing by six to eight years of age. Craniosynostosis is a disorder of the premature fusion of one or more of the major sutures in the skull. The premature fusion can have the appearance of abnormally shaped head and at times facial sutures. The overall cause of craniosynostosis is unknown. Craniosynostosis affects 1 in every 2,500 children and occurs equally in males and females [1,2].

Diagnosis

Seeing a pediatrician after a child is born is recommended for all pediatric patients. The pediatrician will routinely monitor the child's head growth at these well-child visits. A diagnosis of craniosynostosis is made based on symptoms (appearance of the skull and facial structures), detailed case history, and clinical evaluation of the shape of the skull. The skull evaluation can be completed through computerized tomography (CT) scanning and magnetic resonance imaging (MRI). A prenatal diagnosis can be determined through ultrasound examination in some cases; however, most diagnoses are given at or after birth [3].

Types of Craniosynostosis

There are four main sutures of the skull: sagittal, coronal, metopic, and lambdoid [1,4]. Each of four major types of craniosynostosis is named after the location of the premature fused suture. Sagittal craniosynostosis is the premature fusion of the sagittal suture; this forces the head to grow long and narrow. This is the most common type of craniosynostosis, occurring in 40-60% of cases and mostly occurs in boys. Coronal craniosynostosis is the premature fusion of

one of the coronal suture. This may give the appearance of a flattened forehead on the affected side and a bulge on the unaffected side. Unicoronal craniosynostosis may also lead to the turning of the nose and elevation of the eye socket on the affected side. Bicoronal craniosynostosis (both coronal sutures prematurely fused), gives the appearance of a short and wide head with the forehead tilted forward. Coronal craniosynostosis affects 20-30% of the cases and occurs mostly in girls. Metopic craniosynostosis affects the "soft spots" at the front of the skull and affects less than 10% of the cases. Metopic craniosynostosis gives the appearance of a triangular shaped forehead and wider posterior skull. Finally, lambdoid craniosynostosis is the rarest type. It affects the back of the head; giving the appearance of a flat head to one side, one ear higher than the other, or the head is tilted to one side [3].

Most common types of craniosynostosis are non-syndromic primary craniosynostosis [1,2,4]. The cause is unknown, but there are assumptions of fetal position in the womb or valproic acid during pregnancy may increase likelihood. Syndromic craniosynostosis is caused by certain genetic syndromes. This type of craniosynostosis is usually secondary in combination of other symptoms accompanied by the genetic syndrome. There may be a family history associated with syndromic craniosynostosis. Each of these four major types can occur singularly or as a combination of one or more.

Treatment

The treatment for most craniosynostosis is surgery. The most beneficial is to have surgery within the first three months. Surgery should be completed by the first year of life at the very least. If left untreated the intracranial pressure can increase and cause significant developmental delays, cognitive impairment, no energy or interest, blindness, eye movement disorders, seizures, or even death.

Communication Disorders

Syndromic craniosynostosis has a more high risk of hearing loss, low verbal skills and dysarthric speech. Hearing loss can vary in its

form; typically conductive or mixed hearing loss due to craniofacial abnormalities. Children with craniosynostosis may be more at risk for reoccurring otitis media. Children with craniosynostosis may have a developmental or intellectual disability due to damage caused by intracranial pressure which can lead to cognitive impairment. Further the craniofacial abnormalities associated with craniosynostosis can cause problems with articulation and forming of words. It is important to review the person's communication ability with them and their support system. Assistive devices or use of sign-language may be useful with this population. It is important to remember that these people may have cognitive impairments and keep in mind that tests and rehabilitation are completed at developmental age appropriate standards.

Craniosynostosis can be devastating to a family. Many children are not diagnosed until after birth and this may be difficult for the family. It is important that communication disorders professionals be aware of the difficulties and sequela associated with this craniofacial abnormality.

My Family Experience with Craniosynostosis

I have always known my younger brother was different; however, my brother was not treated any differently than my other brother and myself. My parents expected him to do his share at his developmental ability. However, this is not the view that much of the rest of the world has about my brother; he was often referred to the delayed brother or people would say "oh I am so sorry" to my parents when they found out about his disability. As I developed into a more professional career the topic of "People First" became more and more widely accepted as the appropriate language when discussing individuals with intellectual and developmental disabilities. This means that when discussing a person, the person should be addressed before the disability; their disability should not be their identifier. It has been difficult for me to both recognize and demonstrate equality for my brother and other people with craniosynostosis.

Social interactions with people with intellectual and developmental disabilities should be similar if not exactly the same as with anyone else; however, this has not always been the case for people when they converse with my brother. I would hope that people would recognize the need for necessary modifications when talking with a person with a disability in order not to offend or come across condescending. I would argue that everyone makes modifications in their communication depending on who they are talking to with or without a disability.

Personally, there was discomfort for people speaking with my brother who were not as comfortable talking to someone with a disability and are not as knowledgeable about this population. This was my perception as my personal experience; my early communicative skills were skewed because of the early exposure I had speaking to, playing with and having someone with an intellectual and developmental disability in my family.

The one thing that continues to bother me is the use of the word "retard" as a descriptor of my brother and in jargon. As I have grown with my education, I have developed more of a voice for the End The "R" Word campaign. I appreciate that people apologize and have some self-recognition, but I am not the population that deserves the apology. My personal views from interacting with individuals with disabilities and the use of the "R" word are that people assume this population does not understand. I believe that the "R" word still exists today because the origin of the word: this population is dumb. As a sister, I am an advocate. As a future health care professional, it is my responsibility that all individuals receive equal care and treatment from one human to another.

I found that the most difficulty thing people have when speaking with my brother is he looks like an adult, but cognitively is not an adult. People need to be aware of his developmental age. There is a fine art in talking with a person at "their level" without coming off condescending. I have found, at least with my brother, the best idea is to assume a little higher in intellectual capabilities rather than lower. I think that working with individuals with disabilities is a trial and error process, and I have had an seen a lot of good and bad experiences. Some the best experiences I have had, and greatest bonds I have created with individuals with disabilities, is when I have been my truest self. No piece of paper, physical characteristic or diagnosis truly defines an individual. I think that in working and interacting with individuals with disabilities it's important to be who you are and not worry about the minor modifications or slip ups. We are human and that will happen. My brother and our family have learned to politely correct people and laugh it off. It is who we are – it is our family.

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