

UNDERSTANDING AN INVISIBLE DISABILITY


CHIARI I MALFORMATION

By

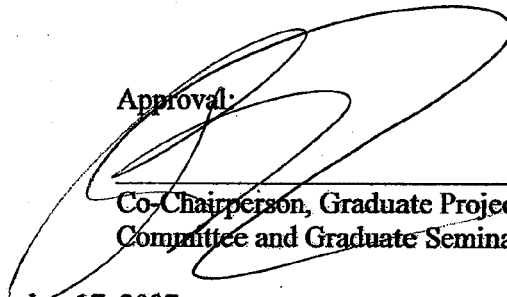
BRIDGET GARRETT

Prepared in partial fulfillment of the requirements of the
Master of Arts Degree in Multicategorical Special Education

Approval:

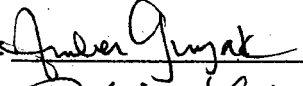

Chairperson, Graduate Project
Committee and Graduate Seminar

Approval:


Co-Chairperson, Graduate Project
Committee and Graduate Seminar

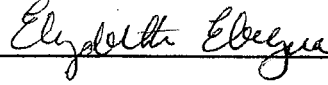
Date: December 17, 2007

Approval of Graduate Seminar Members:

Member: 


Member: 

Member: 

Member: 

Member: 

Member: _____

Member: 

Member: _____

Governors State University

University Park, Illinois

2007

Dedication

I would like to dedicate this final project to my wonderful family who has stood by my side during my journey:

To my dear husband, Matt, I thank you for your patience, your never-ending support and for all of the sleepless nights spent editing this final project.

To my precious son who has been my constant reminder that it is important to take breaks and play. Without play, our lives lack happiness. Thank you, Graham, for reminding me to play.

To my baby girl who is not much of a baby anymore. I began this master's program right after you were born four and a half years ago. Your daily smiles and hugs were just what I needed to keep me going. Thank you, Grace, for your daily dose of sunshine.

Acknowledgements

I would like to acknowledge and thank all of those who assisted me while I worked toward my master's degree at Governors State University. I would like to thank Dr. Maribeth Kasik for her kind heart, empathic ear, and words of encouragement. I would like to thank Mr. Phil Boudreau for his never-ending knowledge of special education. I would also like to thank my panel of peers for all of their support and encouragement. Lastly, I would like to thank my dedicated editor, Matt Garrett, for his eloquent revisions and absolute understanding of this important subject.

TABLE OF CONTENTS

| | |
|---------------------------------------|-----|
| Dedication | i |
| Acknowledgements | ii |
| Table of Contents | iii |
| Title Page | 1 |
| Abstract | 2 |
| CHAPTER 1: INTRODUCTION | 3 |
| Introduction | 3 |
| Statement of the Problem | 4 |
| Purpose of the Study | 4 |
| Questions of the Study | 5 |
| Limitations | 5 |
| Educational Significance of the Study | 5 |
| Definition of Terms | 6 |
| Summary | 9 |
| CHAPTER II: LITERATURE REVIEW | 10 |
| Definition of Chiari I Malformation | 11 |
| Symptoms of Chiari I Malformation | 11 |
| Suboccipital Headaches | 11 |
| Ocular Disturbances | 11 |
| Neurological Disturbances | 12 |
| Anxiety Disorder | 12 |

| | | |
|--|--------|----|
| | Chiari | iv |
| Cranial Nerve, Brain Stem, & Cerebellar Disturbances | 13 | |
| Spinal Cord Function | 13 | |
| Cognitive Difficulties | 14 | |
| Psychogenic Frustrations | 14 | |
| Red Flag Symptoms | 15 | |
| Other Symptoms | 15 | |
| Chiari-Related Disorders | 16 | |
| Syringomyelia | 16 | |
| Hydrocephalus | 16 | |
| Tethered Cord | 16 | |
| Treatment Options | 17 | |
| Surgical Treatments | 17 | |
| Nonsurgical Treatments | 18 | |
| Difficulty of Diagnosis | 18 | |
| CHAPTER III: METHODOLOGY | 19 | |
| Introduction | 19 | |
| Participants | 19 | |
| Instrumentation | 20 | |
| Procedures | 22 | |
| Data Collection | 22 | |
| Data Analysis | 22 | |
| Summary | 23 | |

| | |
|---|----------------|
| CHAPTER IV: DATA COLLECTION AND ANALYSIS | Chiari v 24 |
| Participant Demographics | 24 |
| Questionnaire Results | 25 |
| Background Information | 26 |
| Attendance | 30 |
| Physical Pain | 32 |
| Speech | 43 |
| Attention | 47 |
| Other Relevant Information | 53 |
| Data Analysis | 68 |
| Background Information Analysis | 68 |
| Physical Pain Analysis | 69 |
| Speech and Attention Analysis | 70 |
| Analysis of Other Relevant Information | 71 |
| Summary | 74 |
| CHAPTER V: CONCLUSIONS | 76 |
| Conclusions and Discussions | 76 |
| Implications and Recommendations | 78 |
| Suggested Accommodations & Modifications to Minimize Physical Pain | 79 |
| Physical Education Modifications and Accommodations | 80 |
| Suggested Accommodations and Modifications to Minimize Cognitive Difficulties | 81 |
| Summary | 82 |
| References | 83 |

Running head: Understanding Chiari I Malformation

Understanding an Invisible Disability

Chiari I Malformation

Bridget Garrett

Prepared in Partial Fulfillment for the Requirement of the Masters of Arts Degree in

Multicategorical Special Education

Governor State University

Fall 2007

Abstract

Through an extensive literature review, symptoms commonly associated with Chiari I Malformation (Chiari) that have the potential to obstruct a student's ability to succeed in an educational environment are identified. To study how these symptoms affect students, twenty-two persons with Chiari were asked to respond to thirty open-ended questions. Responses to the written questionnaire provide insight into the challenges facing students with Chiari and may assist educators in their efforts to develop appropriate modifications and accommodations in Individual Education Plans or Section 504 plans.

Understanding an Invisible Disability: Chiari I Malformation

CHAPTER I

INTRODUCTION

How do schools service students for a disability that is unknown to not only those in the educational field, but those in the medical field too? Prior to the enactment of the Individuals with Disabilities Education Act of 1997 (IDEA), many disabled children were prevented from having successful educational experiences in regular school programs because their disabilities were undetected. For many with disabilities, much has changed since the enactment of this act. However, children with unique or rare disabilities that are not fully understood, or even recognized, by the medical and educational communities, continue to face an uphill battle to obtain proper services that will allow them to succeed in school. Children with Chiari (pronounced Kee-AR'-ee) I Malformations (CMI or Chiari) and their families frequently fall into this category and face unique challenges that must be overcome to ensure that these children are able to maximize their educational opportunities.

Chiari I Malformation is a neurological disorder which afflicts approximately 1 in 1,000 people, causing debilitating headaches, neck pain, weakness and numbness in the limbs, balance problems, visual disturbances, and a host of other symptoms (C & S Patient Education Foundation, 2005). Symptoms present during infancy, but they may also delay until adolescence or adulthood (National Organization for Rare Disorders (NORD), 2005). “Most people with Chiari look and act normal until fatigue sets in, they overdo it physically, or they experience pressure changes” (Curtacci, 2006).

Students with this condition, like all students, are entitled to a free and appropriate education. *Appropriate* is defined as educational services designed to meet the individual education needs of students with a disability as adequately as the needs of nondisabled students are met (Office of Civil Rights, 1999). An Individualized Education Plan (IEP) or Section 504 Plan will ensure that students with Chiari I Malformation receive appropriate accommodations and modifications so that their educational needs are, in fact, met as adequately as nondisabled students. In order to do so, however, parents, educators, doctors, and students must first understand what Chiari is, how it affects a particular student in a classroom environment, and how non-extraordinary measures can be taken to meet the student's individual educational needs.

Statement of the Problem

Research reveals that individuals diagnosed with Chiari face physical and cognitive difficulties that are often unnoticed or misunderstood. These individuals struggle with obtaining the proper treatment and medical care because their symptoms are difficult for others to see and understand. As a result of this, students with Chiari may be deprived of services that meet their individual educational needs as adequately as those provided to nondisabled students.

Purpose of the Study

The purpose of this study is to examine, in detail, symptoms commonly associated with Chiari to determine whether they affect a student's educational experience and if so, what accommodations and modifications are needed to maximize these students' educational opportunities. This examination will be based on questionnaires of students

with Chiari as well as an extensive review of available literature. The questionnaires will be used to identify common themes among the responses and to develop a list of appropriate accommodations and modifications needed to help students with Chiari succeed in an educational setting.

Questions of the Study

The following questions will guide the focus of this study:

1. What symptoms are commonly associated with Chiari?
2. How do these symptoms affect a student's success in school?
3. What accommodations or modifications are needed in the school setting to meet the needs of students with Chiari?

Limitations

With any study, there are limitations. Because the participants come from fifteen different states, the researcher is unable to meet face-to-face with those involved. The second limitation is that all of the participants will self report this medical condition and may be inherently biased because they are interested in the outcome. Finally, this study is limited to the fifteen-week time schedule set by the Multicategorical Special Education Program at Governors State University.

Educational Significance of Study

There are very real adverse consequences to students, teachers, and parents if any student's educational needs are not met, and this remains true for students who suffer from Chiari. Students with Chiari may fail due to their struggle with daily symptoms. Additionally, they may feel frustrated if they perceive that nobody fully understands their

symptoms or even accepts that their symptoms are genuine. In an article written about a teenager with Chiari, the author wrote, “Megan and young people like her have to deal with fellow students, some educators, and even some doctors who do not believe their symptoms are real” (Curtacci, 2006). Parents may feel equally frustrated if their child’s teachers are not equipped to meet or even understand their child’s unique needs. Without a basic understanding of Chiari and its symptoms, teachers cannot adequately address their student’s needs.

Definition of Terms

Brainstem. The stemlike portion of the brain connecting the cerebral hemispheres with the spinal cord (Dorland's illustrated medical dictionary twenty-sixth edition, 1981).

Central Nervous System. The portion of the nervous system consisting of the brain and spinal cord (Dorland's illustrated medical dictionary twenty-sixth edition, 1981).

Cerebellum. The rounded portion of the brain, situated above the brainstem, that controls balance and coordination of movement (Random house webster's college dictionary, 2001).

Cerebrospinal fluid (CSF). A fluid, rich in glucose, that circulates in the brain and the spinal column (Random house webster's college dictionary, 2001).

Cervical Spine. The neck region of the spinal column (Random house webster's college dictionary, 2001).

Disability. A disadvantage or deficiency, especially a physical or mental impairment that prevents or restricts normal achievement (The American Heritage Stedman's Medical Dictionary, 2004).

Dysarthria. Difficulty in articulating words due to disease of the central nervous system (The American Heritage Stedman's Medical Dictionary, 2004).

Dysphagia. Difficulty in swallowing or inability to swallow (The American Heritage Stedman's Medical Dictionary, 2004).

Dizziness. A disorienting sensation such as faintness, light-headedness, or unsteadiness (The American Heritage Stedman's Medical Dictionary, 2004).

Dura. The tough, outermost covering of the brain (Dorland's illustrated medical dictionary twenty-sixth edition, 1981).

Foramen Magnum. The opening of the skull through which the spinal cord emerges (The Chiari Institute, 2006).

Nystagmus. An involuntary rapid movement of the eyeball (Dorland's illustrated medical dictionary twenty-sixth edition, 1981).

Occipital bone. A curved, compound bone at the back of the head forming part of the base of the skull (Random house webster's college dictionary, 2001).

Ocular. Of or relating to the sense of sight (The American Heritage Dictionary of the English Language, Fourth Edition, 2004).

Oscillopsia. The sensation that viewed objects are moving or wavering back and forth (The American Heritage Stedman's Medical Dictionary, 2004).

Other Health Impairment. A special education category that “means having limited strength, vitality, or alertness with respect to the educational environment that is due to chronic or acute health problems” (ISBE).

Posterior fossa. It contains three important neurological components: the brain stem, cranial nerves, and cerebellum. The brain stem contains the nerve centers that control eye movements, feelings and movement of the face, hearing, swallowing, shrugging the shoulders, and movements of the tongue. From these nerve centers run the cranial nerves. The brainstem also has nerve centers that control heart and breathing functions. The cerebellum, attached to the back of the brain stem, regulates coordination and fluidity of movement (Oro, 2003).

Section 504. Section 504 of the *Rehabilitation Act of 1973* protects the rights of individuals with disabilities in programs and activities that receive federal funds. Section 504 provides that: “No otherwise qualified individual with a disability in the United States . . . shall, solely by reason of her or his disability, be excluded from the participation in, be denied the benefits of, or be subjected to discrimination under any program or activity receiving Federal financial assistance. The Section 504 regulation defines a person with a disability as any person who (i) has a physical or mental impairment which substantially limits one or more major life activities, (ii) has a record of such an impairment, or (iii) is regarded as having such an impairment (Office of Civil Rights, 1999).

Symptomatology. The combined symptoms of a disease (Dorland's illustrated medical dictionary twenty-sixth edition, 1981).

Syrinx. A cyst that forms within the spinal cord (The Chiari Institute, 2006).

Tachyarrhythmia. An irregularity in the normal heart rhythm (Dorland's illustrated medical dictionary twenty-sixth edition, 1981).

Tinnitus. Ringing in the ears (Dorland's illustrated medical dictionary twenty-sixth edition, 1981).

Valsalva maneuvers. Forcible exhalation effort against nostrils and a closed mouth (Dorland's illustrated medical dictionary twenty-sixth edition, 1981).

Vertigo. A sensation of irregular or whirling motion, either of oneself or of external objects (The American Heritage Stedman's Medical Dictionary, 2004).

Summary

Children with unique or rare disabilities, like Chiari I Malformation, that are not fully understood or recognized by the medical and educational communities, continue to face an uphill battle to obtain proper services that will allow them to succeed in school. Students with this condition, like all students, are entitled to a free and appropriate education (Office of Civil Rights, 1999). The purpose of this study is to examine, in detail, symptoms commonly associated with Chiari to determine whether they affect a student's educational experience and if so, what accommodations and modifications are needed to maximize these students' educational opportunities in order to provide an appropriate education for these individuals.

CHAPTER II REVIEW OF LITERATURE

Definition of Chiari I Malformation

Chiari Malformation is due to a developmental failure of the brain stem and the upper spine, in the cervical region, with no known cause (National Organization for Rare Disorders, 2005). Many researchers believe that the malformation results from an underdevelopment of the lower part of the skull in which the posterior fossa is too small and does not provide the room needed for the cerebellum and lower part of the brain stem (Oro, 2003). Chiari is characterized by abnormalities in the area where the brain and spinal cord meet that cause part of the cerebellum to protrude through the bottom of the skull (foramen magnum) into the spinal canal and it interferes with the flow of cerebral spinal fluid to and from the brain (NORD, 2005).

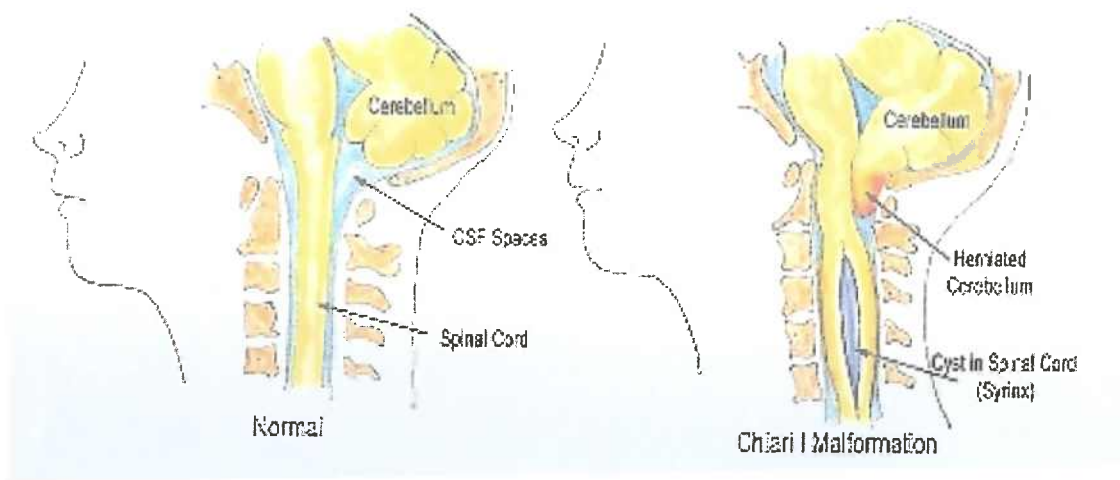


Figure 1. Picture comparing a normal brain to a brain with Chiari I Malformation
From The Chiari Institute Patient Handbook

Symptoms of Chiari I Malformation

The review of literature reveals a complex symptomatology associated with Chiari I Malformation (CMI). The most common constant feature of CMI is a volumetrically small posterior fossa, which predisposes patients to hindbrain overcrowding (Milhorat, 1999). This overcrowding or displacement of PCF contributes to the symptoms. The symptoms develop because of the pressure in the upper cervical spinal canal by the herniation of the cerebellar tonsils down into the cervical canal (The American Association of Neurological Surgeons (AANS) , 2001).

Suboccipital Headaches

The review of literature reveals suboccipital headache as the most common symptom of CMI. This severe headache may possibly be accompanied by neck pain (National Organization for Rare Disorders, 2005). In Milhorat's study of 364 symptomatic Chiari patients from January 1994 to December 1997, these were experienced by 296 patients (81%) and they were described as a heavy, crushing, or pressure-like sensation at the back of the head that radiated to the vertex and behind the eyes and inferiorly to the neck and shoulders (1999). He reports that these headaches are accentuated by physical exertion, Valsalva maneuvers, head dependency, and sudden change in posture (Milhorat, 1999). Garland and Robertson (2001) reported a frequency of 81% of clients with CMI complaining of suboccipital headaches.

Ocular disturbances

Ocular disturbances are disturbances to the sense of sight. Milhorat's study reported that 283 patients (78%) had ocular disturbances (1999). The specific symptoms were

retro-orbital pressure, visual phenomena such as floaters or flashing lights, blurred vision, photophobia, diplopia, and visual field cut and symptoms were also accentuated by physical exertion, Valsalva maneuvers, head dependency, and sudden change in posture (Milhorat, 1999). Pieh and Gottlob describe the cases of two patients. The first patient, with a initial complaint of decreased vision, was reported to have nystagmus on the downward lateral gazes with a bilateral weakness of the rectus muscles and the second patient complained of visual disturbances consisting of blurred vision (2000). Hypersensitivity to bright lights has also been reported (AANS, 2006).

Neurological disturbances

Not only does physical exertion, Valsalva maneuvers, head dependency, and sudden change in posture cause headaches and ocular disturbances, but also neurological disturbances. Seventy-four percent of the patients in Milhorat's study (1999) experienced symptoms of dizziness, disequilibrium, pressure in the ears, tinnitus, decreased hearing loss, vertigo, and oscillopsia. Garland and Robertson (2001) determined a frequency of 57% of patients with a complaint of dizziness. After sneezing, a nineteen year old male presented with persistent numbness and burning in his right shoulder, face and neck, had an MRI and was diagnosed with Chiari (Strayer, 2001). Disorders involving the cerebellum, like Chiari, can include unsteady gait, balance problems, and difficulty with fine motor tasks (Oro, 2003).

Anxiety Disorder

A study of a 34 year old mother revealed panic disorder associated with Chiari I Malformation. She experienced episodes of dizziness, hand tremors, palpitations, chest

