

CONQUER CHIARI®



CHIARI 1000

RECENT ANALYSES AND FUTURE DIRECTIONS

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WHAT IS THE CHIARI 1000?

- Comprehensive self-report survey + Medical Imaging
 - <https://chiari1000.uakron.edu/>
 - Launched September 2015
 - Uses the Internet to gather information from a large number of Chiari patients, both adults and children.
 - Goal= 1000 surveys + 1000 Medical Images (MR, CT)
 - Secure- All identifying information retained separate from all data
- Information collected is used to study various aspects of Chiari malformation and its impact

WHAT IS THE CHIARI1000?



1. Chiari patient completes online Chiari1000 survey and shares MR images to the CCRC
2. Participant is mailed a specially designed t-shirt
3. Data is analyzed
4. Chiari1000 is gateway to future participation in research with the CCRC



WHAT IS THE CHIARI1000?

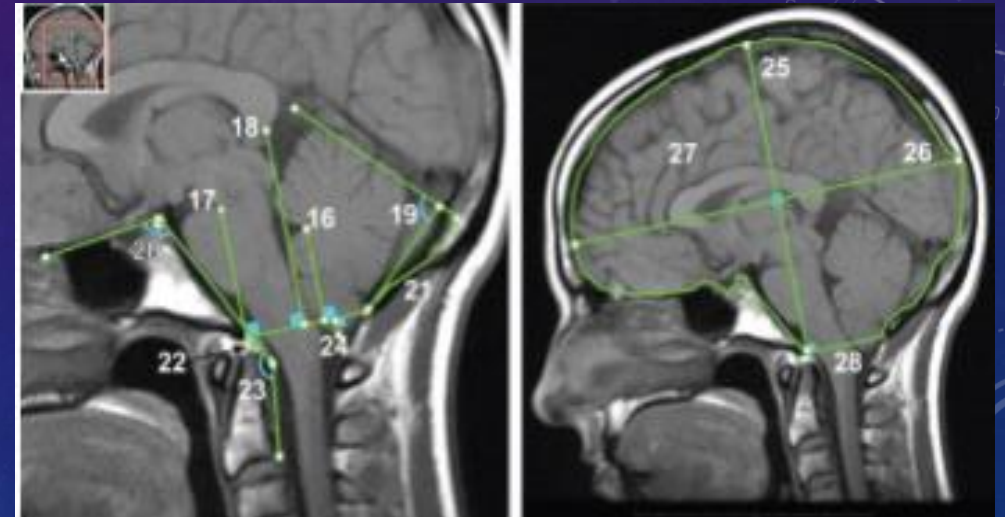
- **Survey Data**
- ~250 questions on:
 - Demographics
 - Chiari diagnosis
 - Symptoms
 - Additional Diagnoses
 - Surgical History
 - Quality of Life
 - Neuropsych variables
 - Depression
 - Anxiety
 - Memory
 - Coping
 - Pain
 - Hypermobility

Analysis



Morphometric Data

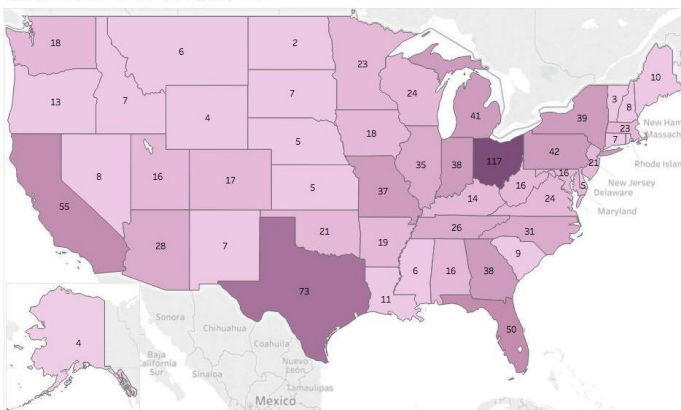
Detailed measurements on MR brain images



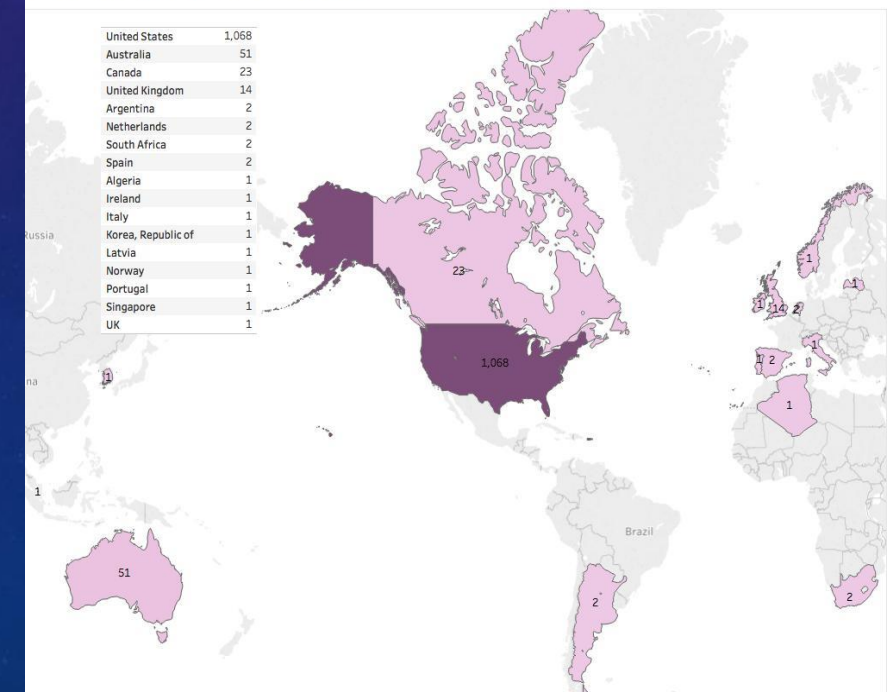
DEMOGRAPHIC

- 1196 People completed the entire Chiari 1000 survey including additional measures
 - 1062 female participants (89%)
 - 134 male participants (11%) (of these only 73 are adult males)
 - 95% Caucasian
- Age-ranges
 - 65 participants- ages 0-7 (5%)
 - 67 participants- ages 8-17 (6%)
 - 1064 participants- ages 18+ (89%)
- Participants from every state (except Hawaii) and 17 different countries

Completed Chiari1000 surveys by state



Completed Chiari1000 surveys by country



FAMILY HISTORY

- Diagnosed immediate family
 - 172 individuals had at least 1 immediate family member with CM
- Diagnosed extended family
 - 136 individuals had at least 1 extended family member with CM

How many immediate family members with Chiari?

| | |
|-----------|--------|
| 1 member | 73.84% |
| 2 members | 19.19% |
| 3 members | 4.65% |
| 4 members | 2.33% |

How many extended family members with Chiari?

| | |
|-----------|--------|
| 1 member | 79.23% |
| 2 members | 16.92% |
| 3 members | 3.08% |
| 4 members | 0.77% |

DIAGNOSTIC HISTORY

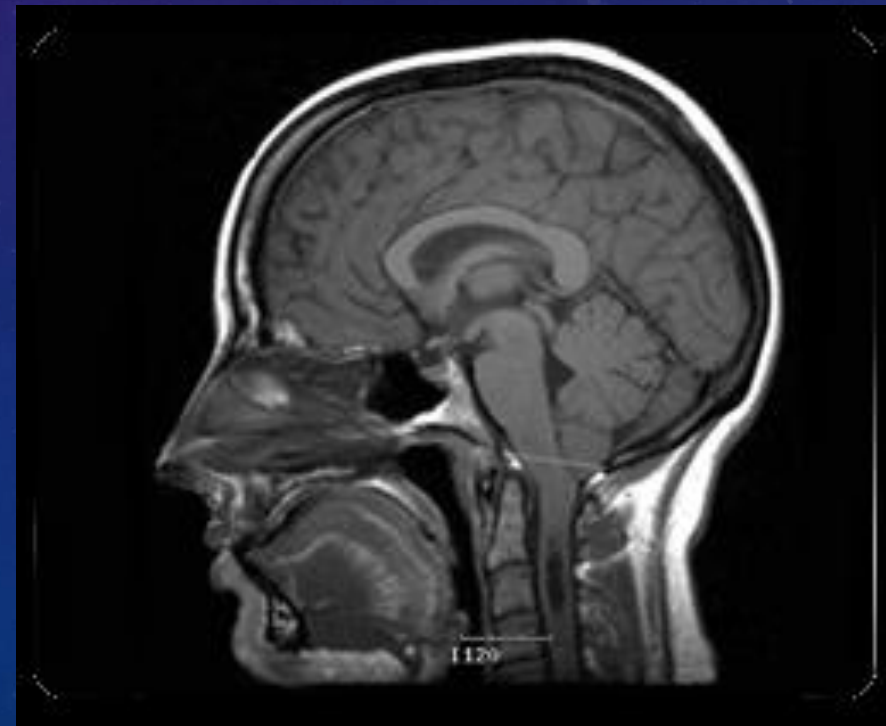
- Age at diagnosis
 - 119 0-7
 - 147 8-17
 - 1,136 18+
- Misdiagnosed
 - 908 (64%) people, who have completed the Chiari 1000, have experienced a misdiagnosis
- Multiple Misdiagnoses
 - 695 (49%) people, who have completed the Chiari 1000, have experienced more than 1 misdiagnosis

ADDITIONAL DIAGNOSES/RELATED CONDITIONS

- Top 10 additional diagnoses/related conditions
 - Migraine headaches (66%)
 - Autoimmune Conditions (37%)
 - Spinal Defects (35%)
 - Scoliosis (22%)
 - Tissue Disorders (21%)
 - Syringomyelia (21%)
 - Fibromyalgia (20%)
 - Chronic Fatigue (16%)
 - ADHD (10%)
 - Ehler-Danlos Syndrome (9%)

GENDER DIFFERENCES IN ADDITIONAL DIAGNOSES

- Men have higher prevalence of
 - Syringomyelia
 - Reynaud's
- Women have a higher prevalence of
 - EDS
 - Migraine Headaches
 - Fibromyalgia
 - Meniere's Disease

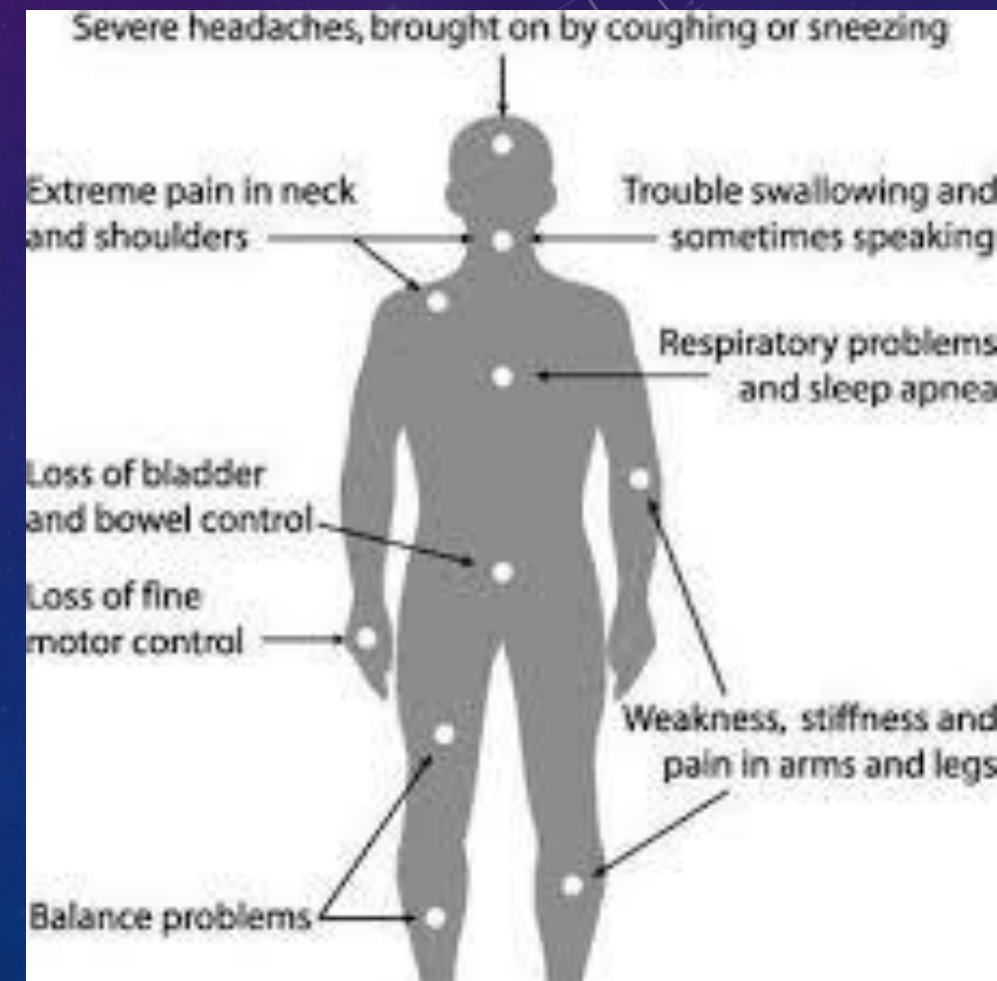


SYMPTOMS

- Top 10 most prevalent symptoms associated with Chiari
 - Headache (93%)
 - Overall Pain (90%)
 - Sleep Interference (81%)
 - Overall Numbness (80%)
 - Neck Pain (79%)
 - Fatigue (75%)
 - Dizziness (74%)
 - Weakness (72%)
 - Balance problems (70%)
 - Shoulder Pain (66%)

SYMPTOM SEVERITY

- Not at all severe 2%
- Mild 7%
- Moderate 32%
- Severe 41%
- Very Severe 18%



GENDER DIFFERENCES IN SYMPTOMS

- Men have higher prevalence of
 - Sleep Apnea
- Women have higher prevalence of
 - Pain
 - Numbness
 - Cognitive issues
 - Weakness
 - Depression
 - Fainting
 - Hearing loss
- No difference in symptom severity despite differences in symptom presentation

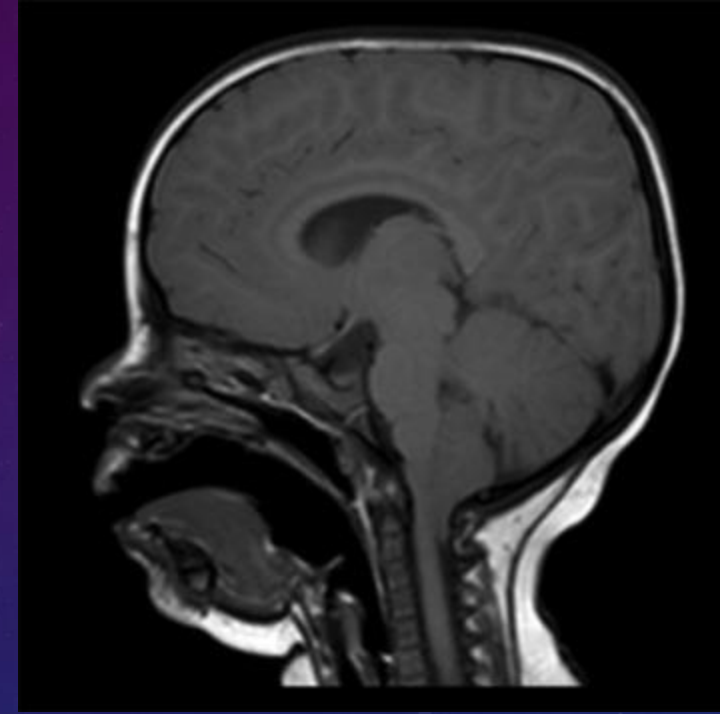
SURGICAL HISTORY

- 766 individuals had surgery related to CM (54%)
- 733 individuals had decompression surgery (52%)
 - 123/733 had multiple decompression surgeries (17%)
 - Ranging between 2-10 decompressions
- Majority of individuals reported at least some improvement after surgery
 - 73%
- No gender differences in surgical history or in surgical impact
 - Despite differences in symptoms

QUALITY OF LIFE CHANGES

- How does CM influence the individual's quality of life?
- 5% of individuals report they cannot work due to CM
- 27% of individuals reported they worked prior to CM diagnosis but stopped after diagnosis
- 46% of individuals report negative financial outcomes related to CM
 - CM was limiting their financial ability
- 43% of individuals reported CM impacting their education/their school experiences

PEDIATRIC CHIARI



- It is important to understand how symptoms, additional diagnoses, and outcomes are represented in children.
- 171 Pediatric cases

DIAGNOSTIC HISTORY

- Age at diagnosis
 - 109 (64%) 0-7
 - 62 (36%) 8-17
- Misdiagnosed
 - 100 (58%) people, who have completed the Chiari 1000, have experienced a misdiagnosis
- Multiple Misdiagnoses
 - 76 (44%) people, who have completed the Chiari 1000, have experienced more than 1 misdiagnosis

ADDITIONAL DIAGNOSES

- Top 10 Additional Diagnoses for Children
 - Migraine Headaches (47%)
 - Developmental Delays (37%)
 - Spinal Defects (26%)
 - Syringomyelia (25%)
 - Tissue Disorders (24%)
 - Autoimmune Disorders (21%)
 - Scoliosis (18%)
 - ADHD (16%)
 - Seizures (15%)
 - Failure to Thrive (13%)

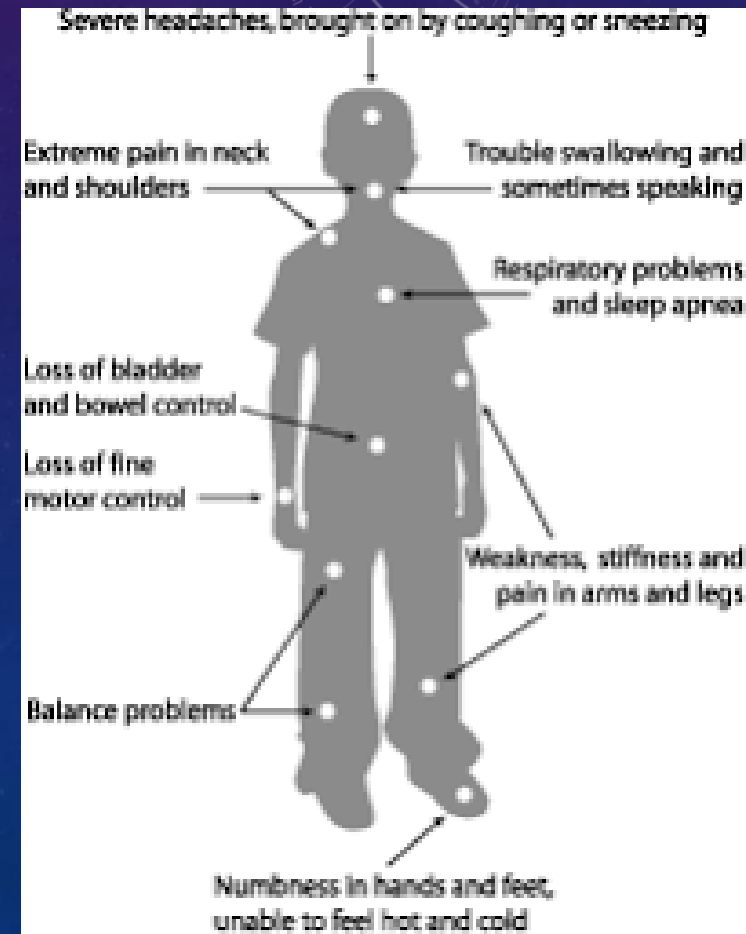
SYMPTOMS

- Top 10 symptoms associated with Pediatric Chiari
 - Dizziness (89%)
 - Headache (80%)
 - Vision Issues (72%)
 - Pain (72%)
 - Cognitive Issues (65%)
 - Balance Problems (55%)
 - Weakness (54%)
 - Fatigue (51%)
 - Sensitivity to Noise (50%)
 - Neck Pain (49%)



SYMPTOM SEVERITY

- Not at All Severe 8%
- Mild 11%
- Moderate 37%
- Severe 33%
- Very Severe 11%



SURGICAL HISTORY

- 84 individuals had surgery related to CM (53%)
- 78 individuals had decompression surgery (93%)
 - 11/78 had multiple decompression surgeries (14%)
 - Ranging between 2-4 decompressions
- Majority of individuals reported at least some improvement after surgery
 - 79%

QUALITY OF LIFE CHANGES

- 19% Withdrew from school
- 54% Reported a negative school impact
- 41% Reported a negative impact on performance at school

PUBLICATIONS AND CURRENT PROJECTS

- There are currently X publications through Conquer Chiari
- Cover a wide variety of topics related to the illness experience
 - Cognition
 - Morphometric information
 - Related conditions
- Upcoming projects
 - Self-report papers for both children and adults using Chiari 1000 survey data
 - DTI
 - Many more!

FUTURE DIRECTIONS

- Because data contains no identifying information it can be shared with researchers outside of the CCRC allowing for a variety of perspectives and analyses
- Identify factors that contribute better/worse surgical outcomes
- Further understand gender differences, age differences, in CM
- Provide researchers and medical professionals with the information needed to reduce potential misdiagnoses