



CCRC Open House 2018: Untangling the Chiari Knot

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In the 4th century B.C., an oracle declared that anyone who could untangle the Gordian knot would go on to rule Asia.



Alexander the Great cut the Gordian knot with a single strike of his sword...

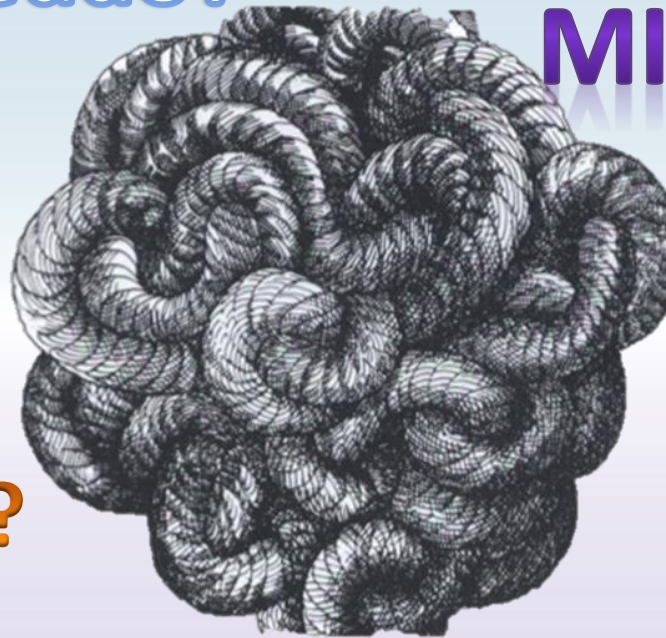
What Causes Symptoms?

What About
EDS, SM, Pseudo?

What Is Chiari?

What Tests
Are Best?

Who Should
Have Surgery?



MISDIAGNOSES!

Pediatric vs
Adult

Quality of
Life

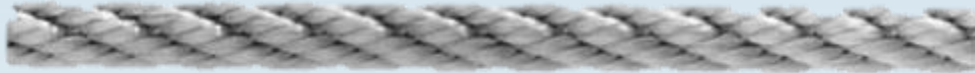
*Chiari is like the legendary Gordian knot,
But we can not cut it with a sword like Alexander did...*

We must untangle it one thread at a time:

Pathophysiology



Symptoms



Diagnosis



Treatment



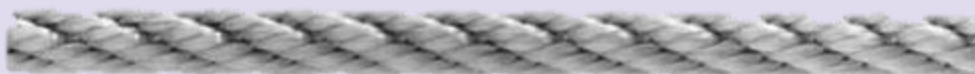
Outcomes



Quality of Life



Related Conditions



State of Chiari: The Challenge

Pathophysiology

No Disease Model
Several Theories

Symptoms

Varied, Diverse, Overlapping
Physical Causes Not Clear

Diagnosis

No Single Definitive Test
TP Not A Good Marker

Quality of Life

Residual Symptoms
Impact Not Known

Treatment

Who Will Respond To Surgery
Traumatic Procedure

Related Conditions

Are All CMs Alike
How Do They Link To Chiari

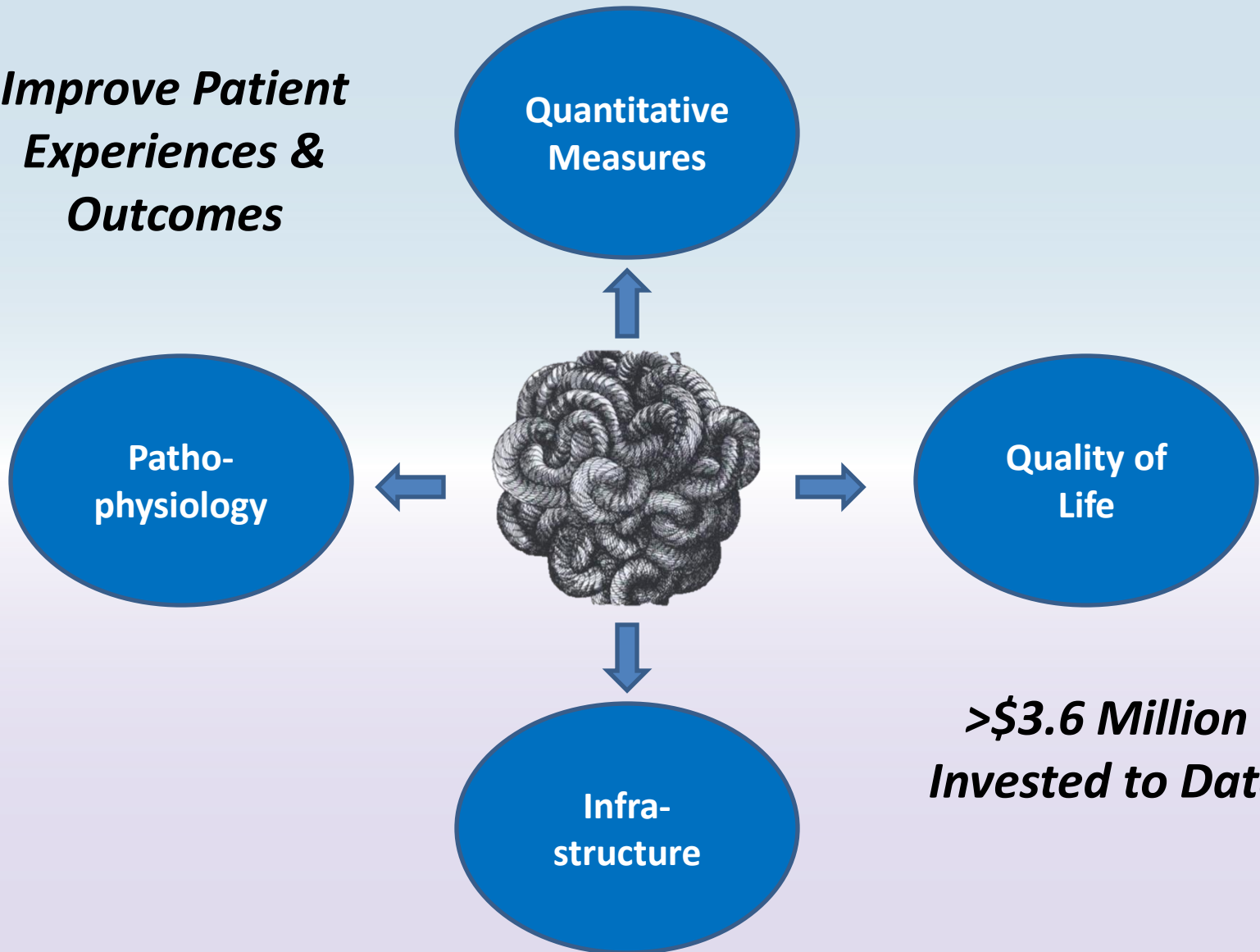
Outcomes

How To Objectively Assess
Adult Outcomes Not Great



Conquer Chiari Research Strategy

*Improve Patient
Experiences &
Outcomes*



Quantitative
Measures

Patho-
physiology

Quality of
Life

Infra-
structure

*>\$3.6 Million
Invested to Date*

Goals

Quantitative Measures

Provide clinicians with quantitative data in order to: assess disease severity; identify candidates for surgery; predict and measure surgical outcome; evaluate new treatments; and classify patients into sub-groups.

Pathophysiology

Develop a complete, practical, and accurate disease model of Chiari, which would answer fundamental questions, enable the categorization of patients, and suggest new techniques for diagnosis and treatment.

Quality of Life

Minimize the impact of Chiari on patients and their families, by maximizing patients' physical, mental, emotional, vocational, recreational, and interpersonal quality of life.

Infrastructure

Develop the research infrastructure necessary to create a vibrant Chiari research community, including: easy access to patient data; cross-disciplinary projects; leveraging 21st century technologies; quickly and broadly disseminating results.

Projects

Quantitative Measures

Morphometrics
Cognitive Impact
Pressure & Strain
Biomarkers
CSF Analysis
Longitudinal Impedance

Pathophysiology

Morphometrics
Genetics
Cognitive Impact
Animal Models
Syrinx Analysis

Quality of Life

Educational Impact
Vocational Impact
Psychological Impact
ACT Therapy
Aquatic Therapy

Infrastructure

Research Center (CCRC)
Chiari 1000
Software Platforms
Apps
Conferences

Conquer Chiari Research Center

Staff

- Executive Director: Loth
- Deputy Director: Allen
- 10 professors, 8 departments
- 2 Full-time post-docs
- 1 Project manager
- 8 PhD students
- 3 Undergrad students



- **\$2.9M Total Funding**
- **\$1M Active Projects**
- **\$.5M NIH Grant**
- **36 Journal Publications**

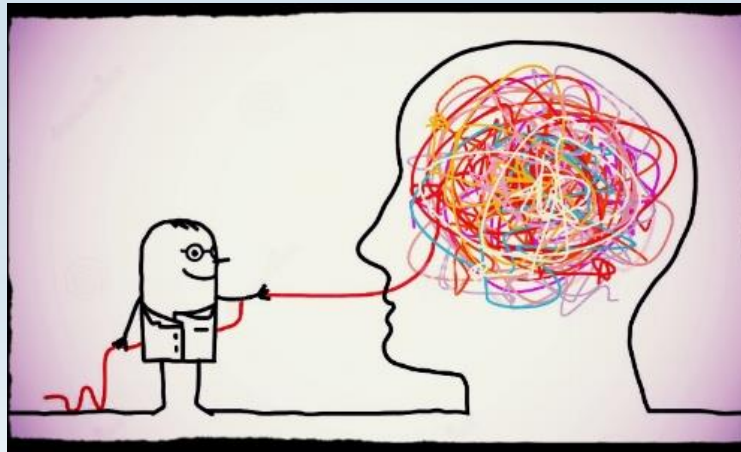
Top 10 Findings To Date From CCRC - (Not Ranked)

1. Up to half of Chiari adults have cognitive issues
2. Advanced imaging (DTI) shows damage to the cerebellum and frontal cortex
3. Emotional impact of Chiari is severe with high rates of clinical depression and anxiety
4. Anatomically, many differences found beyond tonsillar position; entire hindbrain seems to sag
5. Clivus bone is dramatically different, three dimensionally, not just shorter

Top 10 Findings To Date From CCRC - (Not Ranked)

6. Some evidence that the cerebellum is being stretched by an active process
7. Advanced imaging (DENSE) shows areas of extremely high levels of strain in the brainstem and cerebellum
8. Patient perceived outcomes (adults) not as good as clinically published
9. Very high rates of autoimmune diagnoses among adult women – even for diseases with markers, such as Lupus
10. Both ACT and aquatic therapy show promise in helping patients with sub-optimal outcomes

Future Directions



QUANTITATIVE MEASURES

Morph for pediatric & men
Link to symptoms/outcomes
Develop cogn protocol for CM
Automate morph measurements

QUALITY OF LIFE

Develop ACT protocol for CM
Quantify vocational impact
Develop ways to treat SM

PATHOPHYSIOLOGY

Why is clivus bone different?
What role do CTDs play?
Identify specific genes
Explore autoimmune issue

INFRASTRUCTURE

C1000 follow-up surveys
Mobile app development
Continue CCRC growth



Research Is Expensive
Research Takes Time



We Need Your Help...



CYCLE 4 CHIARI

CHIARI 1000
Will You Raise Your Voice?



Get Involved!!