Chiari Malformations
Kerry R. Crone, M.D.
Professor of Neurosurgery and Pediatrics
University of Cincinnati College of Medicine
University of Cincinnati Medical Center
Cincinnati Children’s Hospital Medical Center
Cincinnati, Ohio, USA
kerry.crone@cchmc.org

Objectives
- Define Chiari Malformations
- Explain the differences among the various types of malformations.
- Review the presenting symptoms for Chiari malformations.
- Discuss the options for treatment.
- Recall what you learn today.

Google
- About 35% of U.S. adults say they have used the Internet to figure out what medical condition they or someone else might have.
- A new survey from the Pew Research Center in Washington, D.C. reports that:
  - Among adults who use the Internet to get any kind of health information, 59% admit to diagnostic sleuthing.

From the Internet
- Chiari malformations are named for Hans Chiari, an Austrian pathologist, who first identified types I-III in 1891.
- Julius Arnold further expanded the definition of Chiari malformation type II.

Summary Information
- Some medical sources began using the name Arnold-Chiari malformation.
- Nowadays, some medical sources use Arnold-Chiari malformation as a broad term for all forms.

Summary Information
- Chiari malformations have also been known as:
  - Congenital tonsillar herniation
  - Tonsillar ectopia
  - Tonsillar descent
The Chiari Description

- 1891-Dr. Hans Chiari
- Austrian pathologist
- 17yo female with hydrocephalus who died from typhoid fever and at autopsy her brain displayed an elongation of the tonsils and medial parts of the inferior lobes of the cerebellum

The Arnold Description

- 1894-Dr. Julius Arnold
- German Anatomist
- Portrayed an infant with spina bifida and described an elongated inferior portion of the cerebellum (vermis) that covered the fourth ventricle and extended into the spinal canal.

Synthesized Terminology

- 1907 Schwalbe and Gredig first applied the Arnold-Chiari eponym to patients previously characterized as having the Chiari type 2 malformation.
- Their differentiation was an improper modification of Chiari’s original description. This eponym has continued into modern times to define rhombencephalon deformities.

Why the Difference is Important!

Chiari Malformations

- Chiari I Malformation (Most common)
- Chiari II (Arnold-Chiari) Malformation
- Chiari III Malformation
- Chiari IV Malformation
- Acquired Chiari Malformation
- Chiari 1.5 Malformation
**Anatomic Findings in Chiari I**
- Tonsillar herniation
- Hydrocephalus
- Related Anomalies
  - Anatomic
  - Metabolic
  - Genetic

**Anatomic Findings in Chiari II**
- Herniation of vermis, brainstem and fourth ventricle
- Myelomeningocele
- Syringomyelia
- Hydrocephalus

**Chiari III Malformation**
- Posterior fossa encephalocele that contains cerebellar and brainstem tissue with herniation through an upper cervical spina bifida.
- Grave prognosis.
- Must distinguish this malformation from high cervical myelomeningoceles which may have a favorable prognosis.

**Chiari IV Malformation**
- Absent hindbrain herniation
- Cerebellar hypoplasia or aplasia
- Minimal function

**Acquired Chiari Malformation**
- Spinal Fluid Diversion
  - Development of craniocerebral disproportion related to chronic cranial diversion of cerebrospinal fluid. Essentially "The skull becomes to small for the brain."
  - Herniation of cerebellar tonsils either from chronic lumbar spinal fluid drainage related to lumboperitoneal shunts or acutely related to lumbar external drains

**Chiari 1.5 Malformation**
- Transitional form of Chiari malformation where both tonsils and brainstem are caudally descended into the cervical spine.
- Myelomeningocele is absent.
**MR Findings in Chiari I**
- Herniation of cerebellar tonsils
- Absent intracranial mass lesion
- Tonsillar tip configuration
- Cine MR

**Chiari I Symptoms**
- Pain
  - Occipital-cervical (lower head and neck) pain
  - Back, shoulder, and limb (arm) pain
- Clumsiness
- Dysphagia (difficulty swallowing)
- Dysarthria (difficulty speaking)
- “Dys” means bad, painful or disordered

**Chiari I Clinical Findings**
- Oscillopsia
- Esotropia
  - Crossed Eyes
- Bradycardia
  - Slow Heart Rate
- Apnea
  - Central
- Hoarseness
- Choking
- Gagging
- Hiccoughs

**The “Three” Chiari I Syndromes**
- Brainstem Syndrome
- Spinal Cord Syndrome
- Cerebellar Syndrome

**Brainstem Syndrome**
- Usually from birth through early years
  - Respiratory irregularities
  - Nystagmus
  - Lower cranial nerve dysfunction
  - Recurrent aspiration
  - Pneumonia
  - Reactive airway disease
**Spinal Cord Syndrome**
- Late childhood through second decade
  - Headache
  - Scoliosis (spinal curvature)
  - Motor/sensory losses
  - Hyporeflexia
  - Hyperreflexia

**Cerebellar Syndrome**
- Usually from 18 months through adult life
  - Truncal and appendicular ataxia
  - Sensory motor disturbances

**Radiographic Studies**
- Magnetic Resonance Imaging
  - Pathological hindbrain herniation defined as a distance greater than two standard deviations beyond the range of normal.
    - 6mm First Decade
    - 5mm Second and third decade
    - 4mm Fourth through eighth decade

**Dysphagia**
- Dysphagia is disordered or impaired swallowing
  - Neuromotor dysfunction
  - Developmental disabilities

**Phases of Swallowing**
- Oral Phase
- Pharyngeal Phase
- Esophageal Phase
The Gag Reflex and Vomiting

- The gag reflex is a normal defense mechanism that prevents foreign bodies from entering the trachea (airway), pharynx, or larynx.

Physiology of Gagging

Diagnosis

- History
- Physical Exam
- Radiological Studies
  - MR Scan
    - Head
    - Spine
  - Sleep Studies (apnea)
  - Swallowing Studies (aspiration)

Treatment

- There are only two options in the treatment of Chiari malformations
  - Observation
    - Most common in asymptomatic patients
    - Limited published data
  - Surgery
    - Non uniform approaches

Surgical Treatment

- Bony decompression
  - Foramen magnum (Bone opening skull base)
  - Lamina of first cervical ring
- Dural augmentation
- Tonsillar coagulation
- Fourth ventricle to subarachnoid stent
Surgical Treatment

Posterior Fossa Anatomy

Bony Decompression

Dural Band Incision

Dural Band Decompression

Tonsillar Pistoning
Intraoperative Ultrasound

Dural Augmentation

Intraoperative Appearance

CCHMC Chiari Database
- Over 1200 patients referred since 1992
- 450 patients have undergone surgical treatment
- Temporary Complications < 2.0%
- 90% Patients demonstrated Improvement
- 5% Presenting symptoms stabilized
- 5% No Change

Summary
- Defined the Chiari Malformation
- Described the differences in malformation types.
- Related presenting signs and symptoms.
- Discussed options for treatment.
- Provided an overview of surgery.

My gratitude for attention today.