

THE PEDIATRIC JOURNEY



Chiari I Malformation: Diagnosis and Treatment

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Introduction: The Austrian pathologist, Hans Chiari, published a report in 1891 describing a condition in which the “peg-like elongation of the cerebellar tonsils... accompany the medulla oblongata into the spinal canal.” This condition was not well understood for a number of years until the advent of the brain MRI allowed radiologists and neurosurgeons to better describe and understand the condition. In and of itself, it is not a disease or an illness. Simply stated, it is a mismatch in the growth of the posterior fossa's cerebellum and skull surrounding it. A simple analogy is having size nine feet and wearing size seven shoes. Both the feet and the shoes are normal but the mismatch in size will eventually lead to swollen, tender and painful feet and the inability to walk. One can think of the brainstem and cerebellum as the narrow areas of the two funnels joined together. All of the information from the brain going to the body and vice versa passes through a narrow, small region which when compressed, can lead to a number of seemingly perplexing and unrelated symptoms, making the diagnosis extremely difficult and challenging (Figure 1).

Depending on the severity of the mismatch,

symptoms can develop as early as a few days of life (Chiari crisis) or as late as the fifth or sixth decade of life. Most patients are asymptomatic until an event occurs which causes the symptoms to appear, such as a fall, hit on the head or a car accident. Terminology has evolved since the original paper in 1891. Currently, it is agreed that Chiari I refers to the herniation of the cerebellar tonsils below the foramen magnum in an otherwise normal person. The same term applies to whether the herniation is 1 or 2 mm or as much as 20 mm. Chiari II refers to the presence of the same condition on a patient with spina bifida or a myelomeningocele. Chiari III refers to the herniation of the cerebellum/brainstem into the cervical spine as seen in Figure 4.

Diagnosis: Diagnosis of a Chiari I Malformation can be challenging on a clinical basis as the symptoms can be many and seem unrelated. However, a brain MRI will clearly show the cerebellar herniation through the foramen magnum (Figures 2 & 3). The sine qua non presentation is a headache localized primarily to the back of the head and upper neck. Most importantly, it is a tussive headache, which means that it worsens with Valsalva-like maneuvers such as bending over, coughing, sneezing, laughing, straining or jumping. Tables I-V list a number of other symptoms which can affect the patient, and as previously stated, may seem unrelated to a clinician not familiar with this condition.

Syringomyelia commonly presents in this patient population (Table VI) (Figure 5). For definitive diagnosis, an MRI of the cervical and thoracic spine should be obtained to rule out this condition. A CSF flow study MRI



CEO MESSAGE

As we continue to celebrate all the successes that our medical staff has achieved these last 10 years, we look forward to the next chapter of El Paso Children's Hospital. With the expansion of many service lines, clinics and personnel, we are truly setting ourselves apart to keep more than 85% of our community's kids at home for proper pediatric healthcare.

Our staff and patients are so grateful for the hard work and dedication each one of you has provided to our El Paso Children's Hospital community. This is only the beginning of our next chapter and we are so excited to have you all on this journey with us!

also aids in the diagnosis by demonstrating blockage of CSF from the cranial to the spinal compartments.

Treatment: A significant number of patients are found incidentally to have Chiari I Malformation, yet they are totally asymptomatic. In this group population, if a syrinx is ruled out, no intervention is required and the patients are simply followed clinically. The next group of patients will experience intermittent headaches, which do not affect the activities of daily living (ADL's). They can be managed with analgesics and rest. If the patients presents with progressive symptoms which are severe, uncontrolled

with analgesics and affect ADLs, then surgery is recommended. The presence of a syringomyelia is also an indication for surgery.

The recommended surgery for definitive treatment of this condition is a decompressive posterior fossa craniectomy (removal of the skull on the back of the head), C1 laminectomy opening the dura, lysis of arachnoid membrane adhesions. Reduction/shrinkage of the lower cerebellar tonsils, placement of a dural patch and re-establishment of CSF flow through the base of the skull between the head and spine. (Figure 6)

Outcomes: The surgical treatment of

Chiari I Malformation is associated with great results, outcomes, and minimal complications. Although it is a complex and invasive surgery, it is well tolerated by the patients. Greater than 95% of the patients have complete resolution of the headaches and neck pain. Often times, it is reported by the parents of these patients that many of the accompanying symptoms fully resolve. Children and adolescents indicate major improvement in school performance, memory, concentration, and overall functioning. Complications can include pseudomeningocele formation, CSF leaks and infections.

Conclusion: Diagnosing Chiari I Malformation from a clinical standpoint

can be difficult and challenging. However if a patient (or friend or family member) describe the classic tussive headache (as previously described), referral to a neurosurgeon is in order for definitive diagnosis and treatment.

You can contact our clinic at 915-242-8402 for assessment and consultation on Chiari Malformation.

Table I Symptoms – Infants

- Difficulty swallowing
- Irritability when being fed
- Excessive drooling
- Weak cry
- Gagging/Vomiting
- Arm weakness
- Stiff neck
- Breathing problems
- Developmental delays
- Inability to gain weight

Table II Pain

- Neck
- Back
- Chest
- Arms
- Legs

Table III Weakness

- Face
- Arms only
- Legs only
- Hemiparesis (half of the body)
- Quadraparesis (all limbs)

Table IV Weakness Sensory Loss

- Arms
- Legs
- Neck
- Trunk
- Hands
- Half of body

Table V Miscellaneous

- Ataxia
- Vertigo
- Diplopia
- Dysphagia
- Dysanthria
- Apnea
- Urinary incontinence
- Paresthesia
- Tinnitus
- Drop attacks
- Vomiting/nausea
- Palpitations

Table VI Syringomyelia

- Muscle weakness/atrophy
- Loss of reflexes
- Loss of sensitivity to pain and temperature
- Stiffness of the back, shoulders, arms and legs
- Bowl and bladder dysfunction
- Scoliosis

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Figure 1

Normal midsagittal MRI shows location of the foramen magnum as shown by the red line, which extends from the front to the back of the foramen.

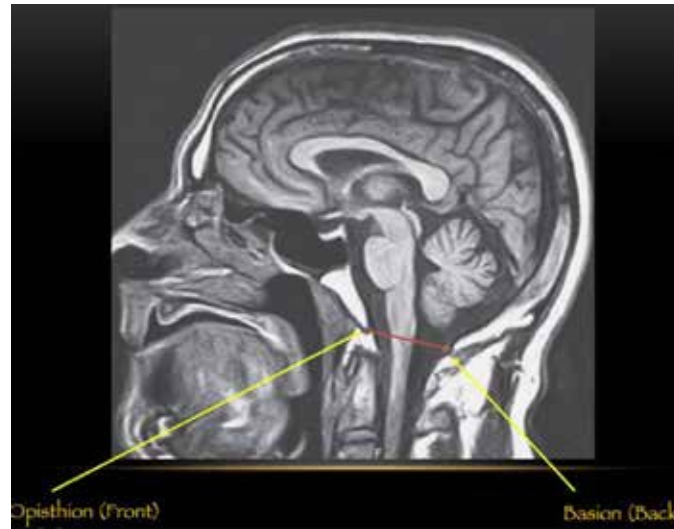


Figure 2

Midsagittal MRI of a patient with Chiari I and cerebella tonsil displacement of 4mm below foramen magnum (red line)

Figure 3

Same as Figure 2 but with displacement of 26 mm below foramen magnum (red line)



Figure 4

Patient with Chiari III malformation showing herniation of the cerebellum into cervical meningocele.

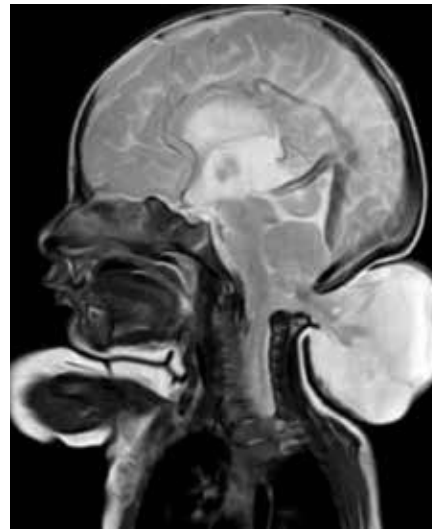


Figure 5

Intraoperative photograph showing reduced right cerebellar tonsil and decompressed brain stem on the right.

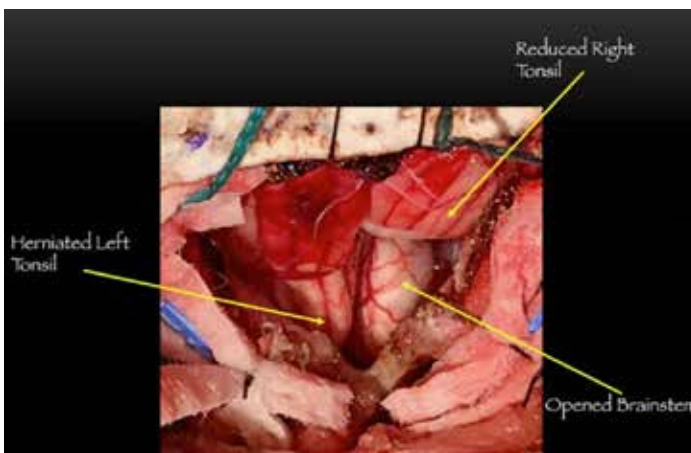


Figure 6

Cervical MRI in a patient with Chiari I malformation and syringomyelia. Fluid build-up within the spinal cord is seen extending between C3 and T1.

CONTACT

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VAX TO THE MAX

Days after the FDA approved the use of the vaccine in individuals under five years, El Paso Children's Hospital administered over 400 Pfizer-BioNTech vaccines to children ranging from six months to four years old.

These first dose vaccines were administered through four onsite clinics on June 23, 2022. The second and third doses will also be offered to patients who received the first dose.

To date over 3,500 children have been vaccinated through El Paso Children's Hospital.



CMO CORNER



Jeffrey Schuster, MD,
Chief Medical
Officer at El Paso
Children's Hospital

I'm extremely proud of our physicians and medical staff for creating a healing and supporting environment here at El Paso Children's Hospital. During this time of growth, it is our responsibility to collaborate and provide the best pediatric care to our community through new expansion projects, such as our recently developed Empower Program.

The Empower Program at El Paso Children's Hospital is a resource for children that are at risk for developing prediabetes or type 2 diabetes, or have been diagnosed with prediabetes or type 2 diabetes.

Our patients and their families can receive nutrition education with follow up appointments to monitor progress. At this location there is also a teaching kitchen, which is used in nutrition education for pediatric patients. On Saturday, June 11, the Pediatric Diabetes Education Services held our first live virtual cooking class in partnership with the Downtown El Paso Lions Club and El Paso Community College Culinary Arts Program.

Preventing Acute Kidney Injury(AKI): Review of Nephrotoxic Drugs

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Acute kidney injury (AKI) is also known as acute renal failure. It is characterized by abrupt decline in kidney function that results in retention of waste products and can lead to fluid overload and electrolyte derangement.

According to Kidney Disease Improving Global Outcome (KDIGO) guidelines, it is defined as increase in serum creatinine by 0.3mg/dl or more within 48 hours or increase in serum creatinine to 1.5 times baseline or more within the last 7 days or urine output less than 0.5 mL/kg/h for 6 hours.

As a pediatric nephrologist, I can recall countless cases of AKI due to nephrotoxic medications and have great concerns due to its high morbidity and mortality in children. AKI with fluid overload and multi-organ injury often require acute dialysis treatment as a bridge to recovery. More importantly, children who suffer an episode of severe AKI are at risk to develop chronic kidney disease (CKD). Due to its negative economic and clinical impact, it is important for us to prevent it.

What are risk factors for AKI due to nephrotoxic agents?

In general, risk factors for nephrotoxicity can be patient related and drug-related.

Patient related risk factors	Drug related risk factors
1. Reduction of kidney function	1. Non-steroidal anti-inflammatory drugs (NSAIDs)
2. Pre-existing conditions	2. Aminoglycosides
a. Cardiac failure	3. Intravenous iodinated contrast
b. Sepsis/critical illness	4. Vancomycin
c. Chronic Kidney Disease	5. Amphotericin
d. Diabetes Mellitus	6. Angiotensin antagonists
e. Volume Depletion	7. Some Anti-viral drugs
f. Low serum albumin	8. Some Anti-cancer drugs.
g. Low serum sodium	
h. Diuretics/Angiotensin antagonists treatment (ACEI/ARB)	

What can we do to prevent it?

There are specific measures based on nephrotoxic medications and general preventive measures.

Non-Steroidal anti-inflammatory drugs (NSAIDs) induced AKI

NSAIDs include Ibuprofen (Motrin), Naproxen, Ketorolac can cause acute kidney injury (AKI). There are multiple types of renal injuries associated with NSAIDs reported in the literature in children; from proteinuria, interstitial nephritis to acute reversible and irreversible renal failure to chronic renal injury. There is no warning labels for the risk for renal injury, or to avoid these drugs in cases of dehydration that can be easily associated in ill children. It is important to ensure adequate hydration and family should be aware of the potential complications.

Aminoglycosides (Gentamicin, Tobramycin)

The major toxic effects of aminoglycosides are ototoxicity (hearing) and nephrotoxicity (kidney injury). Before giving aminoglycosides, we need to assess risk versus benefit. The patients should be given single daily dose and short duration (3 days) of aminoglycoside and need drug level monitoring and measure daily serum creatinine.

Vancomycin

Vancomycin is excreted almost entirely unchanged by the kidney. When kidney function declined, this drug stays in the body for longer hours. It impairs proximal tubular cells function. Careful drug level monitoring and renal function assessment are important in preventing vancomycin induced AKI.

IV Contrast Agent

Contrast induced Nephropathy (CIN) is

usually characterized by 0.5 mg/dl rise in serum creatinine from baseline 24-72 hours following exposure to CT scan with IV radio contrast agent. The only intervention that has consistently been shown to be beneficial is IV hydration.

General Preventive Measures

Before prescribing a potentially nephrotoxic drug, patient's risk factors for nephrotoxicity should be assessed. Modifiable risk factors such as volume or sodium depletion should be corrected. The presence of non-modifiable risk factors should prompt a risk-benefit analysis. If alternative safer drug is available to treat, we should use it. For e.g., Tylenol instead of Motrin for fever.

Correct assessment of kidney function should be done before and at appropriate intervals during treatment to diagnose early kidney injury and make medication dosage adjustment depends on estimated kidney function (GFR). The calculation using serum creatinine level of GFR is $0.413 \times \text{Height (cm)} / \text{serum creatinine (mg/dl)}$. Based on GFR, appropriate renal dose should order. Please see Fig.2 Renal dose based on GFR.

In hospitalized patients, it is also important to determine daily volume status and nephrotoxic medications, review prescribed doses, consider alternatives, and monitor drug level if it is available.

In summary, education, vigilance, and early intervention can prevent nephrotoxic AKI and minimize its impact.

If you need to refer your patients who have history of AKI, presenting with high blood pressure or proteinuria, you can contact Texas Tech Pediatric Nephrology clinic. (Phone no. 915-215 5700 and fax no. 915-215-8694).

Fig. 1 General Preventive Measures

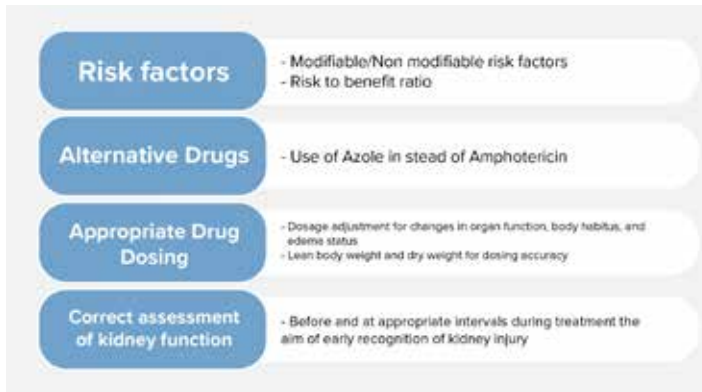
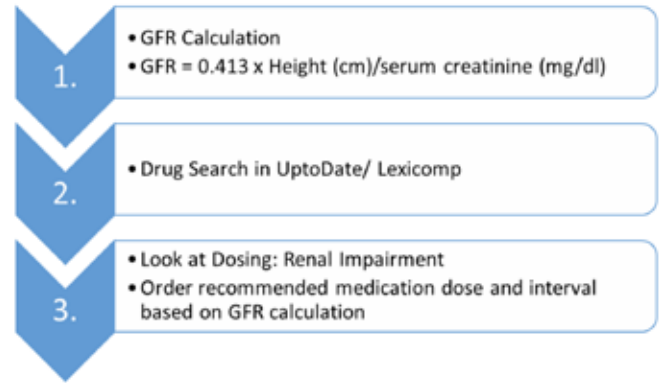


Fig. 2 Renal dose based on GFR.



2022 CME SESSIONS

- Aug 18th | 12:00 PM - Dr. Chiao Topic: Early Childhood Hearing Loss: Opportunities for Care Coordination and Advanced Intervention
- Sep 28th | 12:00 PM - Dr. Bista Childhood Cancer Awareness Month (1 CME)
- Oct TBD | Time TBD - Dr. Diamond Topic Pending (1 CME)
- Oct TBD | Time TBD - Dr. Ibeanu Topic Pending (1 CME)
- Nov 3rd - 5th - Dr. Jimenez Endoscopic Treatments of Craniosynostosis

Congratulations Dr. Chheda



Dr. Sadhana Chheda has been appointed as Vice Chair of the Texas Health and Human Services Commission's Perinatal Advisory Council (PAC).

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CAPACITY FOR CARE

EVERYTHING YOU NEED TO KNOW ABOUT OUR BOND REQUEST



View of UMC's Thomason Tower, Level III.

What is this all about?

The El Paso County Hospital District recently went before the El Paso County Commissioners Court to consider the posting of the District's intent to seek a certificate of obligation bond for \$345.7 million to strengthen the capacity of University Medical Center (UMC) and El Paso Children's Hospital (EPCH). The El Paso County Commissioners Court is scheduled to meet on Sept. 12 to vote on the passage of this request.

What is the need?

University Medical Center and El Paso Children's Hospital both need more beds. For example, UMC often has more critical care patients than available beds. In addition, El Paso Children's successfully reduced the number of children leaving El Paso for necessary care.

UMC and EPCH offer a number of very specialized, unique services in our community that have resulted in an increase in patients who choose to come to these hospitals to address complex health conditions.

A few examples of this type of care include:

- Only Level 1 Trauma Center
- Only Level 1 Comprehensive Stroke Center certified by the Joint Commission
- Level IV Maternal Care (highest level)
- Level IV NICU (highest level)
- Children's Oncology Group

UMC is El Paso's only safety net hospital, meaning that the hospital provides care to all who come to it.

Why now?

While the needs existed before now, the increase in patients and services make capacity needs more urgent at this time.

With the current environment of rapid interest rate increases and rising costs, waiting to address these needs will likely cost taxpayers tens of millions of dollars more. Plus, for the past two years, both hospitals were handling the County's efforts in fighting the COVID-19 pandemic and administering vaccines. It's important to remember that the District has not requested similar funds in nearly a decade.

What will the money be used for?

There are five distinct capacity needs that the District is hoping to remedy via a Certificate of Obligation. These conditions existed prior to COVID-19 but are significantly worse today.

They are:

1. Expansion of Critical Care Beds.
2. Surgery Capacity Care Expansion. UMC needs additional operating rooms and labs necessary for heart and brain care. A special operating room will enable UMC and EPCH to provide care for burn patients who now must leave El Paso and their families for necessary care.
3. A Neighborhood Health and Urgent Care Center in Central El Paso to care for older adults in an area that lacks access to care.
4. Expansion of pediatric beds, operating rooms, and emergency department beds at El Paso Children's Hospital.
5. A Comprehensive Cancer Center. El Paso is one of the largest metropolitan areas in the U.S. that does not have a comprehensive cancer center within a two-hour drive. UMC, EPCH, and Texas Tech University Health Sciences Center El Paso, are best positioned to lead a comprehensive cancer center for El Paso.



Surgery being performed at UMC Main Campus



Construction of the East Tower, funded by the 2007 Bond.

As a taxpayer, what will this cost me?

If the Certificate of Obligation is issued in October of 2022, then you would pay the following for every \$100,000 of your property valuation:

2023 to 2032 - \$56.80 per year or \$4.73 monthly, and then reduced to: 2033 to 2047 - \$28.70 per year or \$2.39 monthly.

Will this produce more jobs for El Paso?

Yes. It is estimated that this will create 400 high paying jobs at UMC and El Paso Children's Hospital, infusing \$31 million into our local economy every year.

In addition, these capacity initiatives will create approximately 850 construction jobs, not including the feeder businesses benefiting from this construction. These building projects will infuse approximately \$38.5 million into our local economy.

We hope these facts help answer your questions about the bond request.

For more information, visit www.CapacityForCare.com or scan the QR code with your phone.

We look forward to expanding our ability to provide world-class healthcare.



THE EL PASO COUNTY HOSPITAL DISTRICT CAPACITY FOR CARE

University Medical Center and El Paso Children's Hospital are asking for \$345.7 million to strengthen their capacity to serve El Paso County.

El Paso, we are working hard to get you all of the facts about our initiative to bring a cancer care center to El Paso and to strengthen our capacity to serve you. But an opposing group is spreading false and inaccurate information to scare people into signing their petitions to

oppose our healthcare work in the region. Don't be misguided. This group is telling you information that is NOT true. There is also strong evidence that those behind these petitions are funded by corporate entities headquartered outside of El Paso.

THE UNTRUTHS THEY ARE TELLING EL PASOANS:

FALSE	FALSE	FALSE	FALSE
That your property taxes are going to rise more than 20 percent as a result of this bond request	That UMC will be the highest taxing entity in Texas.	That UMC is not a good steward of your tax dollars.	That the Hospital District is not being transparent.

ALL 4 STATEMENTS ARE FALSE.

WHAT IS TRUE:

TRUE	TRUE	TRUE	TRUE
Your property taxes will increase by less than 2 percent annually as a result of this proposal.	UMC is one of the smallest of the taxing entities in the County. We are ranked 30th of 39 taxing entities in our region!	The Hospital District dropped its tax rate in 2022 and has dropped it again for 2023.	To date, the District has made 45 community presentations to share the needs, the costs, and explain the details of this bond request - with more meetings scheduled.

We ask: Have those opposed to this been transparent? Who are they? Where are they from? And what is their true motive?

Don't let these people scare you. Don't be fooled or misguided. Get the facts, get the truth, and support the hospitals that support us all.



For more information visit
CapacityForCare.com

