WSCTF
CHIARI MODEL OF CARE

CHIARI MALFORMATION CARE PLAN FOR PRACTICAL USE IN MEDICINE

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Defined by the National Institute of Neurological Disorders and Stroke: “Chiari malformations are structural defects in the cerebellum, the part of the brain that controls balance. When the indented bony space at the lower rear of the skull is smaller than normal, the cerebellum and brain stem can be pushed downward. The pressure that is placed on the cerebellum can block the flow of cerebrospinal fluid (the liquid that surrounds and protects the brain and the spinal cord).”

Chiari malformation can result in widespread symptoms and moderate to severe pain. Symptoms are numerous but typical symptoms reported include headache, dizziness, problems with balance and coordination, visual disturbances, muscle weakness, and numbness.

**Chiari malformation left untreated or poorly managed can cause life-threatening complications.**

Chiari requires a multidisciplinary treatment approach.
THE SEVERITY OF CHIARI IS DETERMINED BY ITS CLASSIFICATION. CURRENT CLASSIFICATIONS THAT ARE RECOGNIZED BY THE NATIONAL INSTITUTE OF NEUROLOGICAL DISORDERS AND STROKE QUICK QUICK SUMMARY:

TYPE I-the cerebellar tonsils descend into the foramen magnum and this does not involve the brainstem.

TYPE II-the cerebellar tonsils and brainstem tissue herniate into the foramen magnum. (May be referred to as Arnold Chiari.)

TYPE III-the cerebellum and brain stem herniate through the foramen magnum and into the spinal cord. Part of the brain’s fourth ventricle may also herniate through the hole and into the spinal cord. In some cases the herniated cerebellar tissue can enter a pouch-like structure that contains brain matter. The covering of the brain or spinal cord can also protrude through an abnormal opening in the back or skull. Type III causes severe neurological defects.

TYPE IV-is identified when the cerebellum is incomplete or underdeveloped. The cerebellar tonsils are in a normal position but parts of the cerebellum are missing.

TYPE 0-no protrusion of the cerebellum but the patient may present with symptoms of Chiari Malformation.
TIME for an action plan:

Chiari Model of Care
PRIMARY CARE PHYSICIAN/PEDIATRICIAN—this is most often the patient’s first contact. Individuals typically present with headaches in the back of the neck or skull …the pain is often worsened by coughing, laughing, or sneezing. Pain may not be relieved with OTC medicines.

**Additional Symptoms:** Patients may present with nystagmus (rapid eye movements back and forth), hoarseness of voice, periods of apnea during sleep, weakness, trouble with balance, abnormal reflexes and there may be additional widespread physical symptoms.

Infants may present with poor suck or feeding and/or failure to thrive. Once testing is completed to rule out traditional reasons for presenting symptoms as determined by the treating physician consider Chiari Malformation. Consider MRI of the brain and cervical spine as the initial test to rule out Chiari Malformation. If MRI is positive for Chiari malformation add MRI of the thoracic and lumbar spine as well to rule out Syringomyelia as this can co-exist with Chiari Malformation and early identification and management is important.

**Neurosurgical consult is recommended as soon as Chiari and/or Syringomyelia is suspected and identified. Do not delay.**

**STEP ONE**
HISTORY-A detailed medical history is important along with a physical assessment. Suspect Chiari Malformation if the child or adult has a history of scoliosis, syringomyelia, myelomeningocele, hydrocephalus, spina bifida, XLH, trauma such as a motor vehicle accident, traumatic brain injury as a veteran, survivors of domestic violence with spinal injury or TBI, premature birth, or genetic background of Chiari or Syringomyelia. **Someone who has a parent or grandparent who was exposed to Agent Orange presenting with symptoms should be evaluated for Chiari Malformation.

MRI Results-if MRI is positive for Syringomyelia or Chiari consult Neurosurgery. Herniations of the cerebellar tonsils both large and small can cause significant complications. Review each case individually considering symptoms on presentation. Someone may have Chiari Malformation and display no symptoms…this is another possibility as well….Chiari could be an incidental finding. Common misdiagnoses are Multiple Sclerosis, Fibromyalgia, Migraines, and at times mental disorders. However, Chiari should be treated seriously.
NEUROSURGERY - determine if the patient with Chiari Malformation has an immediate need for surgical intervention using decompression, tethered cord release, or shunting as a means to stop neurological deterioration and improve symptoms.

HELPFUL TESTING: CINE MRI - special MRI used to evaluate the flow of cerebrospinal fluid; looks at blockage of cerebrospinal fluid as it relates to the Chiari malformation.

Traditional MRI testing initially should include at minimum brain and cervical spine. MRI brain and entire spine is optimal to rule out Syringomyelia as well. The order may be initiated by the primary care physician if a neurosurgical appointment is delayed.

Other tests may include CT scan to see any bony abnormalities. X-ray of the neck in flexion and extension may be used as a simple way to look for any cranio-cervical instability.

STEP TWO
If patient is having symptoms of intracranial hypertension or hydrocephalus such as headaches, dizziness, whooshing in ears, and visual disturbances consider consult with neuro-ophthalmology (see step three)

TREATMENT OPTIONS:
MONITOR—you will be followed closely by your primary care and neurosurgeon and have MRI scans at intervals or for any change in symptoms because surgery may not be needed at the time you are first evaluated.

SURGERY—shunting may be recommended to treat high intracranial pressure that does not respond to oral medications.
Sectioning of the Filum Terminale—minimally invasive surgical technique used to treat Chiari malformation spearheaded in Barcelona Spain.
Posterior Fossa Decompression is a common surgical technique that may be recommended for Chiari malformation when other methods are not recommended.

Post-surgery—Consider if patient is having chronic neurological symptoms (ex: numbness/tingling) consult with Neurology to follow for testing such as EEG, EMG, SSER (somatosensory evoked response) (See step four)

STEP TWO CONTINUED
NEURO-OPTHALMOLOGY—important part of the team for Chiari Malformation to assess the patient for papilledema. Papilledema can lead to blindness if left untreated. Hydrocephalus and intracranial hypertension can co-exist with Chiari malformation and it should be ruled out.

The neuro-ophthalmologist should work closely with the patient’s care team to intervene when necessary to prevent blindness in the children and adults that are affected if symptoms are present that warrant an evaluation as determined by neurosurgery.
NEUROLOGY-important role for medical management. Patients with Chiari Malformation may experience widespread neurological symptoms. Once a neurosurgeon evaluates for surgical intervention neurology can be a helpful provider to help manage neurological symptoms that become chronic. Chiari malformation is complex because of the co-existing complications that accompany it, the adverse affect on the brainstem that can occur as well as spinal nerves that may become damaged causing widespread symptoms with moderate to severe pain.

Neurological Symptoms can include headache, dizziness, nystagmus, difficulty swallowing, visual disturbances, ringing or buzzing in the ears, hearing loss, nausea/vomiting, insomnia, depression, and headaches made worse by coughing and straining. Infants may have irritability when feeding, a weak cry, gagging, vomiting, breathing abnormalities including sleep apnea, stiff neck, and trouble gaining weight.

Helpful testing can include EEG, EMG, BAER, SSEP’S
Chiari malformation requires long term medical management and monitoring over the life-time of the individual who is diagnosed.

Chiari malformation can co-exist with many chronic widespread symptoms as well as disease processes that may require long term medical management. These include Syringomyelia, Hydrocephalus, Autonomic Dysfunction, Cardiovascular Rhythm Disturbances, Central and Obstructive Sleep Apneas, Bowel and Bladder problems, Pain, Spina Bifida, Myelomeningocele, Tethered Cord, Scoliosis, Kyphosis, XLH, and Tarlov Cysts.

Patients who undergo decompression surgery for Chiari malformation may need to undergo more than one surgery in their lifetime.

STEP FIVE
For the remaining specialists required to treat Chiari Malformation the Primary Care Physician and Pediatrician are responsible for making decisions on the appropriate plan of action and specialist to consult depending on the symptoms the patient is experiencing.

**The primary care or pediatrician may initiate an order for MRI if an immediate appointment is not available with neurosurgery. Do not delay MRI scan if Chiari is suspected.**

**Primary Care and Pediatricians please remember that cerebellar herniations both large and small can cause symptoms! Compression of the brainstem can quickly become life-threatening!**

TIME IS BRAIN!

STEP FIVE CONTINUED....
PULMONOLOGIST-Chiari malformation can cause difficulty breathing and changes in respiratory patterns including central and obstructive sleep apneas. Chiari malformation can adversely affect the brainstem.

Useful testing can include PFT’s, Sniff Test, and Sleep Studies.

****Sleep studies are vital on these patients because studies have confirmed that patients with Chiari Malformation have been diagnosed with central, obstructive, and mixed (someone who has a combination of both central and obstructive apneas) sleep apnea. ****

Therapies prescribed could include c-pap, overnight pulse oximetry to monitor oxygen levels, and oxygen therapy.

STEP SIX
CARDIOLOGY has an important role. Syringomyelia, Syringobulbia, and Chiari malformation have been associated with tachycardia, bradycardia, and rhythm abnormalities because or their adverse affect on the brain stem and the spinal nerves as well as the presence of sleep apneas.

Patients with Syringomyelia, Syringobulbia, and Chiari are at risk of developing cardiomyopathy from persistent uncontrolled rapid heart rates from autonomic dysfunction. Sleep apnea is another risk factor and if sleep apnea is left untreated this has already been linked to heart failure.

Helpful testing can include EKG, holter and/or event monitoring, stress test, echocardiogram, and tilt table testing. In patients with a dual family history of cardiovascular complications unrelated to Chiari or Syringomyelia the cardiologist may recommend serial cardiac enzymes (ckmb, troponin) with additional cardiac testing to rule out further complications.

STEP SEVEN
AUTONOMIC DYSFUNCTION SPECIALIST-recommended for individuals with fluctuating orthostatic vital signs and positive tilt table testing to determine a customized treatment plan to treat symptoms of autonomic dysfunction.

**Autonomic dysfunction** can affect temperature, blood pressure, heart rate, and breathing. It is important to develop a customized treatment plan with your doctor and see a specialist to discuss these symptoms to treat symptoms and prevent development of more serious complications long term.
ENDOCRINOLOGY-Growth hormone deficiency has been reported to co-exist in patient cases of Chiari Malformation.

Individuals with known arachnoid cysts have been shown to have hormone abnormalities (estrogen and testosterone production).

Further research is needed in the area of endocrinology and Chiari malformation.
GASTROENTEROLOGY-Consider GI consult for:
-- Persistent symptoms of nausea/vomiting
-- Abdominal pain
-- Constipation
-- Loose Stools
-- Incontinence of bowel

Chiari can cause adversely affect the brain stem and spinal nerves leading to widespread symptoms.

**Straining is not advised for bowel movements so stool softeners are recommended for those with chronic constipation.**

STEP NINE
UROLOGY-Consider consult for urinary frequency and urinary incontinence.

Consider consult for sexual dysfunction.

Chiari can adversely affect the brainstem and spinal nerves leading to widespread symptoms.
OB-GYN-Consider consult for high risk pregnancy for a woman with Chiari malformation depending on the individual symptoms of the patient.

**Folic acid is vital during pregnancy to help prevent neural tube defects.**

Neurosurgery should follow during the labor and delivery and be available to discuss birth plan with the expectant mother and the Ob-Gyn to answer and address any concerns about Chiari and vaginal delivery as well as Chiari and Cesarean section. Currently there is not enough data to support one over the other. The decision is made on a case by case basis.

The neurosurgeon and ob-gyn may go over treatment modalities for pain control customized for you. If the expectant mother has been on long term medication for chronic pain consider consulting in-hospital pain management to adequately address both chronic and acute pain while the patient is in the hospital.
Notify the hospital staff caring for you about your birth plan. Notify your neurosurgeon and care team immediately for any new or worsening neurological symptoms while in labor.

If Chiari is known to be genetic or familial please notify the pediatrician you choose as well as the hospital staff so that any concerns regarding feeding, behavior, and sleep patterns can be monitored beginning at birth for your child.
PAIN MANAGEMENT—a pain specialist that understands the mechanism of Chiari Malformation and the way it can adversely affect the brainstem, spinal nerves, spinal cord, and vessels is important.

The specialist must be willing to learn more about Chiari and its complications. Pain management may be required and can serve an important role in the multidisciplinary treatment plan.

Chiari malformation can cause moderate to severe pain and have a negative impact on the day to day normal activities leading to depression and thoughts of suicide. Pain that is managed adequately can greatly improve the quality of life for the individual & restore some quality of life back to them.

Pain management teams in the hospital should be involved in the management of acute pain in patients who are taking medications for chronic pain at home.

STEP TWELVE
ORTHOPAEDIC SURGEON

Important part of the care team to consult because they are trained in the diagnosis, pre and postoperative management of diseases and injuries of the musculoskeletal system.

Helpful regarding therapy and needed equipment for both pediatric and adult patients with Chiari as well as Syringomyelia because scoliosis, kyphosis, and bony growth abnormalities can co-exist with these diseases.

STEP THIRTEEN
PHYSICAL THERAPY-therapy should be initiated after permission to begin physical therapy has been granted by the neurosurgeon and an order has been obtained.

Physical therapy plan should be re-evaluated and adjusted if needed after each MRI. The physical therapy plan should be discussed and approved by the Neurosurgeon with input from all physicians involved in the patient’s care for a team approach.

The physical therapist should be trained to treat Chiari malformation and understand or be willing to learn about Chiari its co-existing complications. Treatment plans should be customized for each individual.

Our organization is currently working to deliver education to physical therapists about Chiari malformation and its complications.

STEP FOURTEEN
Consider the well-being of the individual living with Chiari Malformation.

Depression can be an ongoing problem with someone who has a chronic illness. It may be helpful to speak to a trained clinician about concerns and ways to cope with the grief of losing health, family, and friends.

We offer three support groups that are open 24 hours a day 365 days a year for individuals living with Chiari Malformation and Syringomyelia. One group is for members, another for caregivers/family, and another for parents and children. We provide resources to assist you while navigating Chiari and Syringomyelia. We encourage you to join!

If you are anyone you know is suicidal or thinking about suicide please call 1-800-273-8255 National Suicide Prevention Lifeline.

STEP FIFTEEN
PALLIATIVE CARE - An important service available to those living with a long term chronic illness.

These services can be ordered by your primary care physician to supplement the care you already receive.

Important to Note: It is not the same as hospice care.

Hospice is often confused with palliative care. They are separate services. Hospice is excellent for end of life care. However, when one has a chronic disease palliative care can be an excellent resource!

Palliative care is a service that would provide additional support to you and your family with resources on ways to cope with ongoing challenges of living with a chronic illness. Palliative care is covered by most insurance plans. Palliative care can be added as part of your medical support team while continuing your active treatment plan with your existing physicians. In addition services provided by palliative care can provide ongoing support to you and your loved ones when you are home from the hospital.

STEP SIXTEEN
DENTAL VISITS-Give a detailed medical history to your dentist and care team if possible prior to the appointment.

MRI’s are an important part of your ongoing care. Make sure the dentist is made aware that any appliance or hardware that is not MRI safe will have to be substituted for hardware that is safe for MRI.

Avoid hyperextending your neck for any prolonged period of time. Offer to provide a towel roll or pillow to help support your head.

Frequent breaks may be needed in the event you become dizzy. Avoid suddenly lying the dental chair backwards because of poor tolerance to sudden position changes.

Inform the dentist if you have gagging or impaired swallowing before any procedure.

Inform the dentist about any cardiovascular abnormalities such as tachycardia from autonomic dysfunction prior to any injections.

STEP SEVENTEEN
Prior to any dental procedure inform the dentist if you have Ehlers Danlos Syndrome because it can affect clotting.

If sedation dentistry is offered you should consider having it done in a hospital setting where emergency equipment and its use is routine due to the complexity of symptoms and this is especially true if you have any form of sleep apnea.

Prior to painful procedures if you are already under pain management make sure your dentist speaks to your pain management physician about the best customized plan for your post op care.
NURSING APPROACH-nurses working with patients who have a diagnosis of Chiari should implement a care plan aimed at the holistic care of the patient. Often times these patients are complex and may require multiple specialists to manage symptoms as it can affect the body head to toe.

Compassion is required because Chiari can cause immense suffering if not managed effectively. Care plans needed will depend on the individual patient and body systems affected. Initial care plans to implement for every single patient with Chiari when they present for care are:

Nursing Care Plan for Spinal Cord Injury
Nursing Care Plan for Chronic Pain
CHIARI MALFORMATION IS COMPLEX. Current patient data suggests that large cerebellar herniations may cause few symptoms while small herniations may result in widespread symptomology. This data shows that ALL cerebellar herniations should be considered along with the clinical picture on a case by case basis.

SYRINGOMYELIA IS COMPLEX. Current patient data suggests that large syrinxes may cause few symptoms while small syrinxes can cause widespread symptomology. This data shows that ALL syrinxes should be considered along with the clinical picture on a case by case basis.

** A benign central canal should not be ruled a benign central canal until there has been at least two repeat MRI scans that show no changes or symptoms. A benign central canal will not grow and will not cause symptoms by definition. Please allow Neurosurgery to make the final call based on a full patient assessment and detailed medical history.

STEP NINETEEN
1. NATIONAL INSTITUTE OF NEUROLOGICAL DISORDERS AND STROKE
Chiari Malformation Fact Sheet
www.ninds.nih.gov/disorders/chiari/detail_chiar.htm

2. AMERICAN HEART ASSOCIATION
Sleep Apnea and Heart Disease, Stroke September 16, 2015
http://www.heart.org/HEARTORG/Conditions/More/MyHeartandStrokeNews/Sl eep-Apnea-and-Heart-Disease-Stroke_UCM_441857_Article.jsp#.V_VCWOArLIU

3. PUB-MED
MRI findings and sleep apnea in children with Chiari I malformation
PMID: 23498564

4. PUB-MED
Haaland Stone WJ, Ittner EA, Teitelbaum BA, Messner LV
Progressive asymptomatic papilledema as the presenting sign of a Chiari I malformation 2012 Mar 30; 83 (3)
PMID: 23231409

RESOURCES
5. MEDSCAPE  Chiari Malformation by Peyman Pakzaban, MD; Brian H Kopell, MD May 13, 2016

6. PUB-MED
Choi, CK and Tyagaraj, K Epub 2013, March 27
Combined spinal-epidural analgesia for laboring parturient with Arnold-Chiari Type I malformation: A case report and a review of the literature
PMID: 23606989

7. PUB-MED
Selmi F, Davies KG, Weeks RD 1995; 9 (4)
Type I Chiari deformity presenting with profound sinus bradycardia: case report and literature review
PMID: 7576283

RESOURCES CONTINUED
8. **PUB-MED**
Stovner LJ, Kruszewski P, Shen JM 1993 Jul-Aug; 8 (4)
Sinus arrythmia and pupil size in Chiari I Malformation: evidence of autonomic dysfunction
**PMID:** 8314116

9. **PUB-MED**
Scoliosis and Chiari Malformation Type I in children.
**PMID:** 21194283
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Worldwide Syringomyelia & Chiari Task Force
http://www.wstfcure.org
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