

# **WSCTF SYRINGOMYELIA MODEL OF CARE**

SYRINGOMYELIA CARE PLAN FOR  
PRACTICAL USE IN MEDICINE © 2016

Model written by Beth Nguyen RN, CEO

- **Syringomyelia** is a disease that is hallmarked by cysts that form inside the spinal cord and expand. The expansion of the cysts can cause spinal cord and spinal nerve damage leading to widespread complications. **Syringobulbia** is a term used when Syringomyelia affects the brainstem. This disease has caused paralysis and death.

**Time for an action plan  
to improve outcomes!**

- **PRIMARY CARE PHYSICIAN/PEDIATRICIAN**-this is most often the patient's first contact. Individuals typically present with complaints of neck and/or back pain unrelieved with OTC medicines. They may also present with sporadic widespread physical symptoms. Once testing is completed to rule out traditional reasons for the back/neck pain as determined by the physician consider Syringomyelia. If patient symptoms persist consider MRI of the brain and spine to rule out Syringomyelia.
- **DETAILED MEDICAL HISTORY**-A detailed medical history is important along with a physical assessment. Suspect Syringomyelia if the child or adult has a history of scoliosis, chiari, tarlov cysts, xlh, autonomic dysfunction, meningitis, myelomeningocele, hydrocephalus, spina bifida, trauma such as an MVA or domestic violence with traumatic brain injury or injury to the spine, genetic background positive for Chiari Malformation, or family history suspicious for Syringomyelia.
- MRI-positive for Syringomyelia or Chiari—consult Neurosurgery.

# STEP ONE

- **NEUROSURGERY**—determine if the patient with Syringomyelia has Chiari or tethered cord and if so determine need for surgical intervention. Surgical approaches can include shunting, decompression, or filum terminale as a means to stop progression and improve symptoms.
- The neurosurgeon will order serial MRI's to monitor patient over time and sooner if new symptoms arise.
- Consider at this time if the patient is having symptoms of intracranial hypertension such as headaches, dizziness, whooshing in ears) If so consider consult with neuro-ophthalmology (See step three).
- Post-surgery-Consider if patient is having chronic neurological symptoms (ex: numbness, tingling) consult with neurology as well for EMG study, BAER, SSEP's, and any other appropriate testing. (See step four).

## **STEP TWO**

- **NEURO-OPHTHALMOLOGY**-important part of the team for Syringomyelia to assess for papilledema. Papilledema can lead to blindness and this is commonly associated with intracranial hypertension. IIH is associated with Syringomyelia and Chiari; if suspected it should be ruled out. Vision loss is important to catch early on or it may become permanent.
- The neuro-ophthalmologist should work closely with the neurosurgeon and primary care physician/pediatrician to intervene when necessary to prevent blindness in the children and adults that are affected.

## **STEP THREE**

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- **NEUROLOGY**-important role for medical management. In a patient who is not a surgical candidate consider the syrinx as the cause of symptoms whether large or small. Syringomyelia is complex because of the potential for spinal cord, and spinal nerve damage causing widespread symptoms with moderate to severe pain.
- Neurological symptoms can include headache, dizziness, rapid eye movement, atrophy of tongue with difficulty in swallowing and speech, numbness, tingling, and burning to extremities, difficulty walking, and loss of fine motor skills. Paralysis can occur. Progression of symptoms can be prevented with early intervention and prompt referral back to neurosurgery for an evaluation. Medical management is important!

## **STEP FOUR**

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- **PRIMARY CARE PHYSICIAN/PEDIATRICIAN-LONG TERM MEDICAL MANAGEMENT.** Syringomyelia has many widespread symptoms that require long term medical management.
- For the remaining specialists required to treat Syringomyelia the Primary Care physician and Pediatrician are responsible for making decisions on the appropriate plan of action and specialist consulted depending on the symptoms. We have already covered the most obvious physicians who have been utilized in the past. However, these are some you may not have considered. Consider step 6!
- **\*\*Primary Care and Pediatricians please remember that syrinxes large and small can cause symptoms! In addition, a syrinx that is unchanged does not mean it is stable. Syringomyelia is a disease and ongoing monitoring is required. A benign central canal is only identified when it is unchanged after more than one MRI and does not cause the patient symptoms. A central canal that is causing symptoms and changing in size is a syrinx and requires evaluation by neurosurgery.\*\***

# STEP FIVE

# 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 Syringomyelia as well as Chiari because scoliosis, kyphosis, and bony growth abnormalities can co-exist with these diseases.

## STEP SIX

- **PULMONOLOGIST**-Syringomyelia can cause difficulty breathing especially if the individual has syringobulbia which is a syrinx high in the cervical spine. Difficulty breathing can also occur when thoracic syringomyelia is present with autonomic dysfunction. Consider consulting pulmonology to follow. Cases of heart failure, heart disease, and pulmonary hypertension have been reported with Syringomyelia.
- Helpful testing can include PFT's, ABG at rest and with exertion, and oxygen therapy may be helpful for some individuals.
- \*\*Sleep studies are vital on these patients as well because cases of central and obstructive sleep apnea have been reported with Syringomyelia and Syringobulbia. Untreated sleep apneas have been linked to heart failure. Central sleep apnea has caused death.\*\*

## STEP SEVEN

- **CARDIOLOGY**—has an integral role. Syringomyelia and Syringomyelia have been linked to autonomic dysfunction, rhythm disturbances, and sleep apneas. Cases of uncontrolled tachycardia, bradycardia, heart failure, and pulmonary hypertension have been reported to co-exist with Syringomyelia alone and with Syringomyelia and Chiari Malformation. If a patient exhibits symptoms of irregular heartbeat, shortness of breath, abnormal blood pressures and heart rates at rest and with exertion, and/or persistent tachycardia please consult cardiology.
- Patients with Syringomyelia and Syringobulbia are also at risk of developing cardiomyopathy from persistent uncontrolled rapid heart rates from dysautonomia.
- Helpful testing can include EKG, holter monitoring, echocardiogram, stress test, orthostatic vital signs, and tilt table testing. If the patient has a strong family history of cardiovascular complications cardiac enzymes can be useful such as CKMB and Troponin levels. If your cardiologist determines a patient is at risk another test may include cardiac catheterization.

## **STEP EIGHT**

- **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.

## **STEP NINE**

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- **ENDOCRINOLOGY**—abnormal hormone levels have been reported to co-exist with Syringomyelia.
- Arachnoid cysts have been found to co-exist with Syringomyelia and arachnoid cysts have been medically shown to increase problems with estrogen and testosterone production.
- There is much more to research regarding the endocrine system and Syringomyelia.

## **STEP TEN**

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- **GASTROENTEROLOGY**—consider GI for persistent symptoms of nausea, vomiting, constipation or diarrhea. Many patients with Syringomyelia have complications related to spinal cord and spinal nerve damage.
- In addition patients should not strain for bowel movements so stool softeners are recommended for those with chronic constipation

# STEP ELEVEN

- **UROLOGY**-consider this consult for difficulty with urinating such as difficulty starting/stopping flow of urine or urinary incontinence.
- Neurogenic bladder has been reported to occur with Syringomyelia.
- Consider sexual dysfunction.

## **STEP TWELVE**

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- **OBSTETRICIAN-GYNECOLOGY**-consider consult for high risk pregnancy for a woman with Syringomyelia.
- Current data does not strongly support vaginal or cesarean section.
- Folic acid is vital during pregnancy to prevent additional neural tube defects.
- Ongoing care for the woman with a diagnosis of Syringomyelia while pregnant should be supervised by the OB-gyn working together with the neurosurgeon to make decisions to customize a birth plan taking into consideration the patient's existing diagnoses.
- Anesthesia Pain management should be consulted prior to labor and birth to develop a plan for management of pain. This is vital if the patient has been on chronic pain medications prior to delivery.
- If concerns are familial Syringomyelia the pediatrician chosen should be notified to monitor the child for any signs of the disease so that early intervention can begin.

## **STEP THIRTEEN**

- **PAIN MANAGEMENT**- a pain specialist who knows about Syringomyelia should be consulted throughout management of this disease.
- Syringomyelia can cause moderate to severe pain that in many cases becomes chronic. This will require long-term management that should be supervised by anesthesia pain management.
- A specialized approach is required.

# STEP FOURTEEN

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- **PHYSICAL THERAPY**-therapy should be performed under the guidance and collaborative effort of all physicians involved in the care of the patient.
- The therapist should be trained to treat Syringomyelia or be willing to learn about Syringomyelia. Therapy should be initiated to prevent further weakness from this disease taking all the patient's individual diagnoses into account.
- Our organization is working with orthopedics and physical therapy to develop a specialized plan of care for adults and children with Syringomyelia!

## **STEP FIFTEEN**

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- **PSYCHOLOGY**-consider the well-being of the individual with the disease.
- Depression can be an ongoing problem with someone who has a chronic illness. Specific to Syringomyelia depression may occur from lack of validation, misdiagnoses, or symptoms that remain poorly managed due to lack of understanding about rare disease.
- 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. The psychologist can help to speak with the care team as well.
- WSCTF support groups are available for you to join others who have the illness that can be helpful as well.
- **National Suicide Prevention Lifeline is 1-800-273-8255**

## **STEP SIXTEEN**

- **PALLIATIVE CARE**-an important approach in dealing with a long term chronic disease is palliative care.
- These services can be ordered by your primary care physician to add to the care you already receive. \*\*It is not 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!
- It is a service that would provide additional support to you and your family on ways to cope with 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 can provide support to your family members and ongoing support when you are home from the hospital.

## **STEP SEVENTEEN**

- **NURSING APPROACH**-nurses working with patients who have a diagnosis of Syringomyelia and/or Syringobulbia should implement a care plan aimed at the holistic care of the patient. Often times these patients are complex and require multiple specialists to manage these diseases as they can affect the body head to toe.
- Compassion is required because this disease can cause immense suffering. Care plans needed will depend on the individual patient and body systems affected.
- Initial care plans to implement for every single patient with Syringomyelia and/or Syringobulbia when they present for care are :
  - Nursing care plan for Spinal Cord Injury
  - Nursing care plan for Chronic Pain

# **STEP EIGHTEEN**

- **DENTAL CONSIDERATIONS:**

- Discuss medical history** and concerns with your dentist prior to starting treatment.

- MRI's are required** over the long haul treatment for ongoing medical management and implants or other appliances that are not MRI friendly should be substituted for appliances that can be used during MRI.

- Voice concerns of dizziness or positioning during treatment.** Lying back in a chair can be uncomfortable and you may need to take breaks during treatment.

- Trouble swallowing should be addressed.** Suction may be needed more often.

# **STEP NINETEEN**

**--EDS can co-exist with Syringomyelia and EDS has in a few cases caused clotting/bleeding complications.**

Discuss this with your dentist prior to any procedure.

**--A diagnosis of tachycardias, bradycardias, arrhythmias and sleep apnea should be discussed with the dentist prior to injections.** Your cardiologist, autonomic specialist, and pulmonologist may need be consulted prior to procedures.

**-- Syringomyelia and Syringobulbia can be complicated. Patients should be evaluated on a case by case basis.**

Some patients may need sedation procedures to be performed in a hospital setting where emergency equipment is available and used frequently. **Patients with a complex medical history may need additional monitoring not available in an office setting.**

**STEP NINETEEN CONTINUED**

## 1. NATIONAL INSTITUTE OF NEUROLOGICAL DISORDERS AND STROKE

Syringomyelia Fact Sheet

[http://www.ninds.nih.gov/disorders/syringomyelia/detail\\_syringomyelia.htm](http://www.ninds.nih.gov/disorders/syringomyelia/detail_syringomyelia.htm)

## 2. AMERICAN HEART ASSOCIATION

Sleep Apnea and Heart Disease, Stroke September 16, 2015

[http://www.heart.org/HEARTORG/Conditions/More/MyHeartandStrokeNews/Sleep-Apnea-and-Heart-Disease-Stroke\\_UCM\\_441857\\_Article.jsp#.V\\_VCWOArLIU](http://www.heart.org/HEARTORG/Conditions/More/MyHeartandStrokeNews/Sleep-Apnea-and-Heart-Disease-Stroke_UCM_441857_Article.jsp#.V_VCWOArLIU)

## 3. MERCK MANUAL

Michael Rubin, MDCM

Syrinx of the Spinal Cord or Brain Stem; July 2014

<http://www.merckmanuals.com/professional/neurologic-disorders/spinal-cord-disorders/syrinx-of-the-spinal-cord-or-brain-stem>

## 4. PUB-MED

Khatwa U, Ramqopal S, Mylavarapu A, Prabhu SP, Smith E, Proctor M, Scott M, Pai V, Zarowski M, Kothare SV 2013 Apr, 48 (4)

MRI findings and sleep apnea in children with Chiari I malformation

PMID: 23498564

## 5. 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

## 6. PUB-MED

Martinez-albaladejol, Prochazkova M, Perez-Sebastian I, Bernardino-Cuesta B, Martinez Ferrandez C, Suarez-Traba OM, Budke M, Garcia-Penas 2016 Sep; 63 (5) 193-200

Syringomyelias in paediatrics: A retrospective study of 25 cases.

**PMID:** 27569564

## 7. PUB-MED

Balaratnasingam C, Cringle SJ, Fatehee N, Morgan WH, Yu DY 2011 Oct 12; 1417; 67-76

Comparison of fluctuating and sustained neural pressure perturbations on axonal transport processes in the optic nerve.

**PMID:** 21911211

## 8. 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

## 9. 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

## 10. PUB-MED

Stovner LJ, Kruszewski P, Shen JM 1993 Jul-Aug; 8 (4)

Sinus arrhythmia and pupil size in Chiari I Malformation: evidence of autonomic dysfunction

**PMID:** 8314116

## 11. PUB-MED

J Neurosurg Pediatr. Krieger MD, Falkinstein Y, Bowen VT, McComb JG 2011 Jan; 7 (1)

Scoliosis and Chiari Malformation Type I in children.

**PMID:** 21194283

## 12. PUB-MED

Qin X, Sun W, Xu L, Qiu Y, Zhu Z 2016 Jul 15; 41 (14)

Effectiveness of Selective Thoracic Fusion in the Surgical Treatment of Syringomyelia-Associated Scoliosis; A Case-Control Study with Long-term Follow-Up

**PMID:** 27398797

## 13. PUB-MED

Jankowski PP, Bastrom T, Ciacci JD, Yaszay B, Levy ML, Newton PO

Intraspinal Pathology Associated with Pediatric Scoliosis: A Ten Year Review Analyzing the Effect of Neurosurgery on Scoliosis Curve Progression. Spine (Phila Pa 1976) 2016 Mar 8.

**PMID:** 26967125

# RESOURCES CONTINUED

14. Riedlbauchova, L., Nedelka, T, Schlenker, J

Symptomatic arrhythmias due to Syringomyelia-induced severe autonomic dysfunction. Clin Res Cardiol 2014 Oct; 103 (10): 839-45

**PMID:** 24847769

15. Zizzo M, Lanaia A, Negro A, Santi R, Bonilauri S

Total gastrectomy for rare refractory gastroparesis in patient with Syringomyelia; A good impact on quality of life. Ann Med Surg (Lond) 2015 Oct 30; 4 (4); 444-7.

**PMID:** 26693008

**RESOURCES CONTINUED**



# **WSCTF SYRINGOMYELIA MODEL OF CARE**

Worldwide Syringomyelia & Chiari Task Force

<http://www.wstfcure.org>

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