

**ADDIS ABABA UNIVERSITY
COLLEGE OF HEALTH SCIENCES
SCHOOL OF GRADUATE STUDIES
DEPARTMENT OF RADIOLOGY**



**CROSS SECTIONAL STUDY ON MAGNETIC RESONANCE IMAGE
FINDING OF SYRNIX AND ASSOCIATED LESIONS IN ADDIS ABEBE
UNIVERSITY TIKUR ANBESA SPECIALIED HOSPITAL AND SAINT
POULS MEDICAL MILLINIUM COLLAGE , ADDIS ABABA, ETHIOPIA
FROM SEPTEMBER 2018-SEPTEMBER 2019.**

**INVESTIGATOR: DR. BARGICHO JUHAR (MD, RADIOLOGY
RESIDENT)**

**A RESEARCH REPORT FOR PREPARATION OF SENIOR PAPER TO
BE SUBMITTED TO THE RADIOLOGY DEPARTMENT, COLLEGE OF
HEALTH SCIENCE, ADDIS ABABA UNIVERSITY IN PREPARATION
FOR PARTIAL FULFILLMENT OF THE REQUIREMENTS FOR THE
POST GRADUATE STUDY IN RADIOLOGY.**

**ADDIS ABABA, ETHIOPIA
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INVESTIGATOR; BARGICHO JUHAR .MD, RADIOLOGY RESIDENT

ADVISORS: GETACHEWU ASSEFA, MD, PROFESSOR OF RADIOLOGY, AAU

COLLEGE OF HEALTH SCIENCES, DEPARTMENT OF RADIOLOGY, AA, ETHIOPIA.

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**Addis Ababa, Ethiopia
September , 2011**

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**THIS THESIS IS ACCEPTED IN ITS PRESENT FORM AS SATISFYING
THESIS REQUIREMENT FOR THE STUDY OF SPECIALITY
CERTEFICATE IN RADIOLOGY.**

APPROVED BY THE EXAMINING BOARD

ADVISORS: GETACHEWU ASSEFA, MD, PROFESSOR OF NEURO RADIOLOGY

SIGNATURE _____ DATE _____

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I would like thanks to all who were on my side during the this work specially saint pouls residents and department team I

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Acronym

ACR	American College of Radiology
ASNR	American Society of Neuro-radiology
CSN	Canadian Syringomyelia Network
CT	Computed tomography
MRI	Magneticresonance imaging
NMR	Nuclear magnetic resonance
SCBT-MR	Society of Computed Body Tomography and Magnetic Resonance

Abstracts

Background

Syringomyelia is a disorder involving the spinal cord which is abnormal accumulation of CSF in the spinal cord . Pathologically, it is characterized by the presence of longitudinally oriented cavities and gliosis. The term "hydromyelia" has been used to describe the appearance of dilatation of the central canal of the spinal cord while the term "syringomyelia" has been reserved for cavities independent of the central canal .From a practical viewpoint, it is impossible to differentiate most cases of true hydromyelia from those of true syringomyelia. Consequently, recent literature tends to unite the two terms syringohydromyelia or to use the terms "syringomyelia" or "syrinx," in a generic sense to refer to the spectrum of disease that is involved without implying endorsement of a specific pathogenesis hypothesis

Syringohydromyelia .caused [associated] with the Chiari malformations,traumatic syringomyelia, idiopathic syringomyelia, and syringomyelia associated with tumors and associated with cervical spondilosis

Objective

To determine common site of spinal cord syrinx on clients who has syrinx on MRI

To describe Causes of syringomyelia and associated lesions on pediatric age

To assess syringomyelia and associated lesions on adult

Method

The design of the study will be prospective cohort study

The study will be conducted on patients 50 patients on selected hospital in Addis Ababa on patients who had spinal and if available brain MRI to study associated lesion compared with each and associated lesion

The data collected through organized questionnaire The collected data analyzed using SPSS version 20 software and interpretation of the data will be done

Results

CM-I was the most frequent associated condition (present in 14 patients [28%]), followed by spinal tumor (11[22%]) from which (2[4%]) had associated kyphoscoliosis, post traumatic (4[8%]) from post traumatic (3[6%]) had associated kypho-scoliosis, Chiari II (4[8%]), Chiari III (3[6%]) and only kypho-scoliosis (3[6%]) see table 1. No associated brain or spinal cord condition was found in 3 patients (6%), and syringes in these patients were considered idiopathic. The other diagnoses are (2 [4%]) patients had diastomatelia, two (2[4%]) tethered cord and one kyphosis (1[2%]) and one infectious cause (1[2%]) There are also two patients who had multiple diagnoses kyphoscoliosis with diastomatelia (1[2%]) and kyphoscoliosis with tethered cord at L3 vertebra (1[2%])

Conclusion

Syrinx location within the spinal canal also differed according to associated condition. Syringes associated with CM-I, CM-II, or CM III had a more superior cranial extent, usually in the cervical spine. Syringes associated with tethered cord and spinal dysraphism were more likely to have their cranial and caudal extents located more caudally.

The incidence of kypho scoliosis did not differ significantly by associated condition, with the exception that patients with Chiari malformation were less likely to have scoliosis on our study.

KEY WORDS ,syrinx, Chiari malformation; syringohydromelia

1. Introduction

1.1 Background

1.2 Statement of Problem and Significant of Study

1.1.2.1 Syringohydromyelia

The term “syringomyelia” refers to any cavity within the substance of the spinal cord which may or may not communicate with the central canal. The term “hydromyelia” refers to dilatation of the persistent central canal of the spinal cord which communicates with the fourth ventricle. The wall of the cystic cavity in the central canal in hydromyelia is lined by ependymal tissue where as in case of syringomyelia is not lined by ependymal tissue. The cavity in both lesions contains cerebrospinal-like fluid.[1] The differentiation between hydromyelia and syringomyelia is at times difficult to make by radiological, clinical or pathological methods. There has been much confusion with the use of these terms, as some authors have used these terms interchangeably. Harwood-Nash has suggested the use of the combined term, syringohydromyelia, to avoid confusion.

1.1.3 Classification

Syringohydromyelia may be divided into two broad categories:

1. Primary or congenital

These lesions are associated with developmental anomalies at the foramen magnum and in the posterior fossa most often the Chiari malformation

2. Acquired or secondary syringohydromyelia

- A. Posttraumatic
- B. Post-inflammatory
- C. Associated with spinal cord tumors
- D. Vascular insufficiency

1.1.4 Clinical Features

Syringohydromyelia is frequently associated with congenital anomalies of the neural and spinal axes. These include spinal dysraphism, myelocoele, the Chiari malformation, the Dandy-Walker syndrome, diastomatomyelia, scoliosis, the Klippel-Feil syndrome and spinal segmentation defects. Although syringohydromyelia may present at any age, it is primarily a disease of childhood or early adult life. It is characterized by: a loss of sensation to pain and temperature,

with preservation of light touch and proprioceptive sensation. This reflects the interruption of the decussating fibers mediating pain and temperature by the syrinx; atrophic changes (skin lesions, Charcot's joints); anterior horn cell involvement (muscle weakness with wasting and absent reflexes); upper motor neuron involvement (weakness, spasticity, hyper-reflexia), and pyramidal tract involvement (abnormal plantar reflexes).

Syringomyelia causes neurological deficits because of gradual compression of the spinal cord. Its clinical presentation includes progressive weakness in the upper and or lower extremities, diminished sensation, and chronic pain. People with this disorder are frequently misdiagnosed because of vague signs and symptoms. A delay in the diagnosis of this disorder can result in irreversible neurological deficits. These deficits can be reversed, however, by early and effective decompression of the spinal cord.

A survey of the Canadian Syringomyelia Network (CSN) participants during their 1996 annual meeting[2] discovered a 97.5% prevalence of pain; 47% of the participants were completely unable to work, with the overwhelming majority attributing this to intractable pain.

Of the participants,90% stated that there was a connection between stress, activity, and pain. The most common pain complaints in this survey involved the head/neck region (90%), shoulders/arms (75%),and chest/abdomen (50%).

This survey triggered the present descriptive study in patients with syringomyelia to elucidate:

(A) pain onset and evolution, temporal and spatial characteristics, and severity/interference with quality of life;

(B) The relation of pain and somatosensory abnormalities; and MRI characteristics of cavitory lesions.

The characteristic site of onset of the lesion is at the lower end of the cervical cord, although any part of the cord or the entire cord may be involved.

Syringomyelia is an uncommon disease that is caused most often by

type I Chiari malformation, which develops in the hindbrain, and less frequently by other factors which are not limited to the hindbrain, including trauma, infection, or scoliosis

The patho-physiology of syringomyelia (cavitations]within the spinal cord remains controversial [3–6]. Most cases have been associated with type I Chiari malformation. Others have been associated with typically non congenital conditions, including scoliosis, arachnoiditis, and trauma. However, idiopathic syringomyelia is rare.

Syringomyelia is more easily detected nowadays because of the availability of spinal magnetic resonance imaging . Before this technique became popular, the nonspecific and highly versatile signs and symptoms of syringomyelia – such as chronic pain, hyperhidrosis, hypertension, limb paresthesia, sensory loss, progressive weakness, and, in some cases, ascending paralysis [5, 7] resulted in syringomyelia being easily overlooked, especially in patients with minor signs and symptoms. Consequently, most patients were diagnosed so late in the course of the disease that its neurological sequel were irreversible. Fortunately, the neurological deficits associated with syringomyelia can be reversed if decompression is carried out earlier in the course of the disease.

Current theories about the mechanism for the formation of syringomyelia are controversial. Possible mechanisms include perforation of the foramen of Magendie resulting in the subsequent expansion of the central canal (the Gardner theory) and the “ball-valve” effect of obstruction of the foramen magnum associated with type I Chiari malformations (the Williams theory) [8 ,9].

Obstruction of the cerebrospinal pathway results in a pressure gradient which is relieved when pressure is dissipated through potential spaces.

Eventually, this results in the creation of an intramedullary cavity.

Conditions leading to syringomyelia can develop within the hindbrain or elsewhere [7,9]. The most common hindbrain lesion that results in syringomyelia is type I Chiari malformation, which develops within the foramen magnum [10–11].

Spinal cord trauma, the second leading cause, can also lead to meningeal fibrosis and syringomyelia [5, 7, 12–14]. Other causes of syringomyelia do not necessarily involve the hindbrain such as spinal cord tumor, infection, kyphosis, and a reaction to iopendylate (Pantopaque) [5, 3, 6, 15].

NUCLEAR MAGNETIC RESONANCE

Prior to the advent of computed tomography , especially when used with intra-theccametrizamide, and, more recently, nuclear magnetic resonance ,the diagnosis of syringohydromyelia was difficult and not consistently reliable. These new imaging modalities have revolutionized the diagnosis of syringohydromyelia.

NMR is a new imaging technique that generates high quality medical images which in many cases are more accurate and more specific than existing imaging techniques. Preliminary experience using NMR in the evaluation of syringohydromyelia indicates that NMR appears to be a sensitive and less invasive method for the diagnosis and delineation of syringohydromyelia than other modalities including CTMM. NMR can distinguish relatively pure fluids like cerebrospinal fluid from more inhomogeneous fluid collections. This may make it possible to differentiate between the cavities seen in syringohydromyelia and those seen with neoplasti cyst

The practice parameter was revised collaboratively by the American College of Radiology (ACR), the American Society of Neuro-radiology (ASNR), and the Society of Computed Body Tomography and Magnetic Resonance (SCBT-MR).

Magnetic resonance imaging of the spine is a powerful tool for the evaluation, assessment of severity, and follow-up of diseases of the spine. Spine MRI should be performed only for a valid medical reason. While spinal MRI is one of the most sensitive diagnostic tests for detecting anatomic abnormalities of the spine and adjacent structures, findings may be misleading if not closely correlated with the clinical history, clinical examination, or physiologic tests. Adherence to the following practice parameter will enhance the probability of detecting such abnormalities.

Spine MRI has important attributes that make it valuable in assessing spinal disease. Alternative diagnostic imaging tests include radiography, computed tomography ,myelography, and CT myelography. Compared with these other modalities, MRI does not use ionizing radiation. This is particularly advantageous in the lumbar area where gonad exposure may occur, and in the cervical spine to avoid radiation to the thyroid. Myelography requires an invasive procedure to introduce intra-theccal contrast agents. Both the puncture and the contrast agent can produce side effects and rarely significant adverse reactions. MRI allows direct visualization of the spinal cord, nerve roots, and discs, while their location and morphology can only be inferred on plain

radiography and less completely evaluated on myelography. Compared to CT, MRI provides better soft tissue contrast and the ability to directly image in the sagittal and coronal planes. It is also the only modality for evaluating the internal structure of the cord.

However, MRI has not completely supplanted CT for spine imaging. For example, CT provides better visualization of cortical bone than MRI, and some patients who have contraindications to MRI will require other modalities, usually CT, for primary evaluation. While not a contraindication to spine MRI, metallic hardware in the area of scanning may in some cases limit the usefulness of MRI. In selected cases, more than one of these modalities will be needed for a complete evaluation.

1.1.2.2. INDICATIONS

Indications for spine MRI include, but are not limited to, the evaluation of:

1. Congenital spine and spinal cord malformations
2. Inflammatory/autoimmune disorders
 - a. Demyelinating disease.
 - b. Connective tissue disorders, e.g., systemic lupus erythematosus.
3. Infectious conditions
 - a. Spinal infection, including disk space infection, vertebral osteomyelitis, and epidural abscess.
 - b. Spinal cord infection including abscess.
4. Vascular disorders
 - a. Spinal vascular malformations and/or the cause of occult subarachnoid hemorrhage.
 - b. Spinal cord infarction.
5. Degenerative conditions
 - a. Degenerative disk disease and its sequelae in the lumbar, thoracic, and cervical spine.
 - b. Neurodegenerative disorders such as subacute combined degeneration, spinal muscular atrophy, amyotrophic lateral sclerosis.
6. Trauma
Nature and extent of injury to spinal cord, vertebral column, ligaments, thecal sac, and Paraspinal soft tissues following trauma.

7. Neoplastic abnormalities

- a. Spinal canal tumors .
- b. Intra-Dural extra-modularly masses.
- c. Intramural leptomeningeal disease.
- d. Extra-dural soft tissue and bony neoplasm's.
- e. Treatment fields for radiation therapy.

8. Miscellaneous

- a. Spinal abnormalities associated with scoliosis.
- b. Syringohydromyelia (multiple etiologies, including Chiari malformations, trauma
- c. Postoperative fluid collections and soft tissue changes (extradural and intradural).
- d. Pre-procedure assessment for vertebro-plasty and kypho-plasty.

MRI is essential for diagnosis of syringomyelia. Syringomyelia fluid has the T1- and T2-relaxation characteristics of CSF, and multiplanar imaging allows assessment of the width, dorsal horn involvement and longitudinal extent of the cavity. The shape of the cavity may be complex with septations (haustria) and generally involves a portion of the central canal at some level [2,20]. Syringomyelia, however, is merely the effect of an obstruction within the subarachnoid space, and the goal of imaging is to determine the cause of the syringomyelia. In the instance of the Chiari-like malformation, the basioccipital bone is presumed to be short, resulting in a reduced caudal fossa volume, the caudal cerebellar vermis and the medulla extend into or through the foramen magnum, and there may be ventricular dilatation.

1.1.6 Treatment of Syringomyelia

The treatment strategy for patients with this disorder varies with the extent of disease progression. Some patients show no signs or symptoms during disease progression for many years; such patients may be treated conservatively [7, 10,].

Syringomyelia has been reported to resolve spontaneously with conservative treatment in a few cases but some patients deteriorate progressively with that approach. Such patients may better be treated with surgical decompression, comprising myelotomy, syringe-subarachnoid or syringopleural shunt, and spinal cord transection [7].

There were no previous study on our population

2. LITRATURE REVEIW

The diagnosis of syringomyelia or hydromyelia is made only when the intramedullary cavity is imaged. The spinal cord is often enlarged, but at times it may be smaller than normal [22,31, 35, 36, 41]. The size of the cord may be evaluated by myelography or metrizamide CT, and occasionally by plain CT of the cervical region [23, 30-36].

Intra-medullar cavities are rarely demonstrated on plain CT and often require delayed metrizamide CT examinations [21, 14-16].

A common associated abnormality of hydromyelia is the Chiari I malformation, which has been suggested to play an important part in the pathogenesis of intramedullary cavities

[21 , 22,29, 42-44]. The presence of such an anomaly and ventricular communication of the hydromyelia has an important bearing on surgical therapy.

Ventricular decompression of cysts that do not communicate with the ventricle in the presence of a Chiari I malformation does not ameliorate the clinical symptoms and often makes them worse [21 , 22, 24, 25,27-29,45, 46].

Other causes of syringes such as tumor, radiation therapy, trauma, and hemorrhages have to be sought, as these primary diseases also affect therapy [21 , 23, 44, 45].

Spinal Cord Size Intramedullary cavities are not always associated with enlarged spinal cord. MR demonstrates changes in spinal cord size in the cervical region, but is not as sensitive as myelography or metrizamide CT in revealing slight alterations.

The presence of kyphosis and scoliosis makes it difficult to align MR sections accurately in the thoracic region in a substantial proportion of cases, and results in poor imaging of the thoracic spine. The smaller diameter of head compared with the body coil also permits better resolution in imaging of the upper cervical spinal and cranio-cervical junction than is obtained in the rest of the spinal cord using the body coil. SE 500/30 is the optimal pulse sequence for demonstrating the spinal cord. On the whole, sagittal sections are superior to coronal views.

Evaluation of spinal cord size is by visual impression, as currently the resolution of MR is inadequate for accurate quantitative measurements. It has been suggested that spinal cord size is

dependent on the posture of the patient, namely that it collapses in the upright position [22, 35]. This undoubtedly occurs during air myelography, where there is a difference in hydrostatic pressure of fluid inside an intramedullary cavity and the subarachnoid space [21, 22, 41]. This mechanism cannot be used to explain a small cord size, either on metrizamide studies, plain CT scans, or MR images, where the pressures in these spaces are equal.

Signal of cervical cord cyst is not as low as in uncomplicated cyst. Inhomogeneous signal within cord cavity compatible with mixed solid/cystic components. Homogeneous low signal for comparison. Cavity varies in size in different parts of the spinal cord and atrophy is present in unpredictable regions. Generalized cord atrophy has been a concomitant finding in some cases studied.

Intramedullary Cavities

Intramedullary cavities have been divided into hydromyelia, which is a dilatation of the central canal, often communicating with the ventricular system, and syrinx, arising primarily outside the central canal [21, 22, 34]. The latter occurs secondary to intramedullary tumors, trauma, and bleeding into the spinal cord [21, 23, 45, 46].

Hydromyelia has been thought variously to arise from interference with the ventricular or subarachnoid pressure and pulsation, in the presence of a Chiari I malformation

[21-23, 42-44]. From a therapeutic standpoint, the important factor is to determine whether the cavity communicates with the fourth ventricle and if cerebellar tonsillar ectopia is present.

CT resolution is better in the cervical than thoracic regions:

Large cervical spinal cord cavities are occasionally visible on plain axial CT scans, especially when high-resolution scanners are used [27]. Smaller cavities are demonstrated only when they fill with metrizamide. It is unknown what proportion of cavities are thus filled, even with delayed repeat CT examinations.

The reason MR failed to reveal cavities within small spinal cords may well be the small size of the cavities, which are invisible because of partial-volume effect on the 10-15-mm-thick MR sections.

Preliminary published data using surface coils and stronger magnets indicate substantial improvement in spatial resolution in examinations of anatomic structures close to the surface, such as the orbit and the temporal bones [47, 48]. SE 500/30 imaging seems to be the optimal technique for demonstrating cavities. It is important to image cord cavities on both sagittal and coronal views. The axial section is sometimes useful in viewing the cavities, especially in the thoracic region, where CT scans are considerably degraded by artifacts. However, because of the limited anatomic field covered by multiple-plane axial sections, it is not possible to view the entire spinal cord using a single set of scans; this method should be reserved for diagnosing cavities that are shown inadequately in the other planes.

Filling of an intra-medullary cavity on metrizamide CT does not mean that there is communication with the ventricular system. It has been variously suggested that filling occurs by trans-ependymal/cord passage of contrast material, filling via an open communication with the subarachnoid space at the conus medullaris, or via the fourth ventricle at the obex [35, 36, 49]. The cord cavities may extend into the medulla; some have rounded configurations and appear to enlarge the medulla and distort the fourth ventricle, with which it does not seem to communicate. In other cases the upper margins are tapered, suggesting a small communication with the ventricular system, even though a passage is not seen due to the limited spatial resolution. In other cases the cavities are situated entirely within the cervical and thoracic cord, quite distal to the ventricular opening.

The caudal limit of cavities that end in the cervical region are usually clearly demarcated; those that terminate in the thoracic cord are seldom defined due to the poor resolution of our current MR imaging in this location.

As clear communication between intramedullary cavities and the ventricular system is almost never demonstrated, it is not usually possible to differentiate syringomyelia from hydromyelia. Truly eccentric location within the spinal cord may be more characteristic of syringomyelia than of hydromyelia.

Spinal cord tumors may be partly cystic. The cystic and solid parts of tumors are seen to have different signal intensities; the locations and configurations of the cord expansion may also be eccentric and irregular.

In cases of syrinx associated with a cord tumor the cyst had an appearance identical to simple hydromyelia, but the tumor itself often had an abnormal signal on SE 2000/120 images. The relation of the cavities to the tumor was less clear-cut in the case that had been treated by surgery and radiotherapy. It is possible that the latter was the sole cause of the syrinx, since meningioma is rarely associated with syringomyelia [45].

The incidence of asymptomatic hydromyelia in patients investigated for clinical symptoms of the Chiari I malformation is unknown. The use of MR in the investigation of diverse neurologic disease has already detected some asymptomatic Chiari malformations, and undoubtedly incidental hydromyelia will be revealed. Irrespective of the mechanistic relation of the Chiari malformation to production of hydromyelia, it is generally agreed that it should be treated at the same time as the cord cavity is decompressed [22, 24, 25, 28, 29].

Tonsillar ectopia is detected on sagittal views: the tonsils are sometimes contiguous and inseparable from the posterior margin of the spinal cord, and may be confused with localized tumor expansion of the medulla and cervical cord [50].

For the last 50 years the world literature contains reports of familial syringomyelia only in a brother and sister (Barre and Reys, 1924), in two sisters (van Bogaert, 1934), and once in monozygotic twins (Wild and Behnert, 1964).

Syringomyelia has always been recognized as a non-familial condition. Evidence that the Chiari type I malformation is familial is difficult to find. However, the type II malformation, the more severe form of the condition (Chiari, 1891), is associated with hydrocephalus and meningomyelocele which have an increased familial incidence. An environmental rather than a genetic factor has been postulated as the cause for this because of the low incidence in monozygotic twins.

Tethered cord syndrome is a diverse clinical entity characterized by symptoms and signs which are caused by excessive tension on the spinal cord due to impaired ascent^{51,52} of the spinal cord. This may be secondary to any of a heterogeneous group of disorder e.g spinal lipomas, lipomatous filum, and split cord malformations. It typically occurs in children, but it is a rare pathologic entity in adults. The association of syrinx and tethered cord has been documented and there is a suggestion that syrinx formation can result from tethering leading to neurological

deficit Few cases of TCS coexisting with syrinx have been [53 ,54] reported and findings suggested that the tethered cord preceded the syrinx formation.53 Hsu et al reported a case of syringomyelia with associated aortic coarctation and tethered cord syndrome, where serial imaging showed significant reduction in thoracic syrinx after the repair of the coarctation and release of tethered of cord, indicating a possible cause effect relationship between syngomyelia and tethered cord.

Erkan et al reported thirty two cases of tethered cord with associated terminal synomyelia in the United States of America and noted that the frequency of sacral tethering was 40.6%, with the intramedullary paracentral position of the syrinx being 75%. Our patient had lumbar tethering with a central syrinx. Erkan et al concluded that radiological significant terminal syringomyelia affects the clinical presentation of the tethered cord syndrome. Our patient declined?? surgery, so we couldnot monitor resolution of clinical symptoms and the syrinx.

Management of TCS in adults may involve surgical detethering in those that are asymptomatic and decompression in those with associated syrinx .

Tethered cord association with syringomyelia in adults is rare, although with the use of modern imaging tools like MRI, the diagnosis of TCS is no more as rare as was thought.

There is a need to include TCS as an important differential diagnosis in patients that present with lesion or disease of the spinal cord and MRI is recommended as an importantdiagnostic work-up in thesesome syringes are not associated withnoany of the pathologic conditions and may be considered idiopathic.]55 56,57,58\It is well established that large cervical cord syringes inpatients with typical-appearing CM-I are causally linked in many cases.

Cervical spinal spondylosis is an extremely common condition that has only rarely been described as a cause of syringomyelia. [58–63] T. Milhorat et al. reported 115 cases with non-neoplastic spinal cord syringes and there was only one patient whose syringomyelia was initially caused by cervical spondylosis. [58]

3. Objective of study

3.1 Main objective

To determine associated lesions of syringohydromelia on spinal and brain MRI

3.2 Specific objectives

To determine common site of spinal cord syrinx on clients who has syrinx on MRI

To describe Causes of syringomelia and associated lesions on pediatric age

To assess syringomelia and associated lesions on adult

4. Research methodology and design

4 Materials and Methods

Patient Selection

We reviewed the spine MRI reports over a 12-month period and identified 50 patients 19 female and 31 male whose reports contained one of the following terms: syringomyelia, hydromyelia, or syringohydromyelia.

We reviewed the clinical and imaging data of this study 50 patients met the following inclusion criteria: syrinx was present, FOV included entire syrinx and any associated spinal cord signal intensity abnormality, most had no contrast -enhanced MRI of the entire syrinx (38[78%])some have (12 [22%]) contrast enhanced MRI was performed, and the syrinx was imaged in the axial T2 and sagittal planes on both T1 and T2 weighted imaging

4.1 Study Setting; The study conducted at the saint pouls hospitalsand TikurAnbesa specialized hospital which has spinal MRI ,or spinal with brain MRI and image interpreted by neuro-radiologist, ,radiologist and radiology residents

4.2 Study Design; The design of the study will be prospectivedescriptive study

4.3 Sampling method; The sampling method is non-probability sampling technique which is based on inclusion criteria

Selection of sample size the spinal and or if available spinal and brain MRI who has syrinx

4.4 Sourcepopulation; Patients who has syrinx on spinal, if available brain MRI

4.5 Study population; patients imagewhich has syrinx on spinal cord ,if available brain MRI

4.6 Inclusion and exclusion criteria

4.6.1 Inclusion criteria

- All patients scanned with spinal or , spinal with brain MRI who has syrinx on imaging

4.6.2 Exclusion criteria

Inadequate image quality

Patients who has only brain MRI is not included in this study

4.7 Sampling technique and sample size

All patients with spinal and or if available with brain MRI that has fulfilled the Inclusion criteria

4.8 Data collection plan

Data collected using structured questionnaire from archiving images . The MRI reports will be reviewed and findings recorded in the questionnaires.

4.9 Data quality control

In order to evaluate the clarity of the questionnaire, validity of the instruments and after the pretest, the findings and observations obtained will be used to modify the questionnaire and the data collection process accordingly.

4.10 Data collection Analysis and interpretation of Data

The data collected through organized questionnaire which includes demographic parameters. The questionnaire administered to residents after gaining their consent and information regarding the above parameters entered filled by the trained data collector and radiology residents

Data regularly checked for quality by giving training and supervising The collected data analyzed using SPSS version 25.0 software and interpretation of the data done based on the above stated study parameters

5 Ethical considerations

The research underway after it has been given approval by the Institutional Review Board of the College (IRB). The confidentiality of the information is maintained. The clients' images enrolled in the research got formal standard interpretation and evaluation.

6. Results

A total of 50 patients [100%] had syringes who had MRI with a minimum of 1.8mm and a maximum of 9 mm in maximum axial dimension were identified.

There were 19 female and 31 male patients age from 1 day to 63 years old patient included.

The syringe location relative to center of spinal cord is 45 patients [90%] are central on spinal cord and 5 patients [10%] had off midline with extension to the centrecanal.

CM-I was the most frequent associated condition (present in 14 patients [28%]), followed by spinal tumor (11 [22%]) from which (2 [4%]) had associated kyphoscoliosis, post-traumatic (4 [8%]) from post-traumatic (3 [6%]) had associated kyphoscoliosis, Chiari II (4 [8%]), Chiari III (3 [6%]) and only kyphoscoliosis (3 [6%]) see table 1. No associated brain or spinal cord conditions were found in 3 patients (6%), and syringes in these patients were considered idiopathic. The other diagnoses are (2 [4%]) patients had diastomatelia, two (2 [4%]) tethered cord and one kyphosis (1 [2%]) and one infectious cause (1 [2%]).

There are also two patients who had multiple diagnoses: kyphoscoliosis with diastomatelia (1 [2%]) and kyphoscoliosis with tethered cord at L3 vertebra (1 [2%]) see the pie chart 1. Syringe characteristics differed according to associated condition. In general, syringes associated with CM-I, CM-II, idiopathic syringes and dysraphism were wide compared with tumor (mass).

Syringes in patients with CM-I had a minimum and maximum of 2.3mm and 8mm respectively.

Similar to those in patients with CM-I, Chiari II had a minimum and maximum of 5mm and 8mm respectively.

The post traumatic patients had minimum and maximum 3 mm to 4.5cm respectively which is less than idiopathic which are minimum and maximum 1.8 to 6mm respectively

In contrast to other causes the patient who had tumor/mass has minimum and maximum of 3.1mm to 9 mm average 6.05mm which is largest relative to chiary I, chiary II , chiary III , idiopathic tethered and kyphoscoliosis patients, one patient (1[2%]) had kypho-scoliosis with spinal cord mass [dermoid tumor]

The most consistent chiary I magnetic resonance imaging findings were obliteration of the retrocerebellarcerebrospinal fluid spaces, tonsillarherniation on of at least 5 mm all patients and varying degrees of post fossa anomalies.

Almost all idiopathic syringes were less than or equal to 6 mm in maximum axial dimension (3 syringes [6%]).

The most common age diagnosis of chiary I were 30 years up to 40 years minimum 8 and maximum above 51 where as spinal cord mass had age 15 up to 28 years

The chiary II and chiary III age are below 10 years where as kyphoscoliosis and kyphosis 20 to 30 years and less than one year's respectively

Syrinx location within the spinal canal also differed according to associated condition Syringes associated with CM-I, CM-II, or CM III had a more superior cranial extent, usually in the cervical spine. Syringes associated with tethered cord and spinal dysraphism were more likely to have their cranial and caudal extents located more caudally. Idiopathic syringes had an intermediate position within the spinal canal. CM-I-associated syringes were longer than idiopathic syringes (C1 -T12 level and C3 to T7 level respectively . There are (8 [16%])patients with KyphoScoliosis which is defined as a Cobb angle of at least greater than 10°, there are only KyphoScoliosis found in 3 patients (6%) , spinal cord tumor with kyphoscoliosis (2[4%]) kyphoscoliosis with tethered cord (1[2%]) and kyphoscoliosis with diastomatomeia (1[2%]),

There is only one patient had kyphosis only (1[2%]).

The incidence of kypho scoliosis did not differ significantly by associated condition, with the exception that patients with chiarymalformation were less likely to have scoliosis

We evaluated conus level to rule out occult spinal cord tethering in patients with idiopathic syrinx, as their width morphology was similar to those with tethered cord. We found a normal mean conus position in patients with idiopathic syrinx, in contrast to those with tethered cord and those with spinal dysraphism in whom a low conus position was expected. The mean conus position for patients with CM-I and syrinx was in the normal range. When looking at the cranial extent of syringes, we found that it was in the cervical spine in 100% of patients with CM-I-associated syringes compared with of patients with spinal cord mass associated syringes. A small number of patients with CM-I had narrow syringes with cranial extent in the thoracic or lumbar spine, more similar in morphology to that of an idiopathic syrinx than a typical CM-I-associated syrinx.

Despite these outliers, however, the combination of syrinx width greater than 5 mm and cranial extent in the cervical spine had 99% specificity for CM-I-associated syrinx. Conversely, syrinx width of 5 mm or less and cranial extent in the thoracic or lumbar spine had sensitivity for idiopathic syrinx.

The age at the time of syrinx diagnosis, including patients in all groups, was minimum of one day to 63 years old. Syringes associated with chiary II, tethered cord and spinal dysraphism were diagnosed earlier, with a mean age at the time of diagnosis of 8 years for tethered cord-associated syrinx, and less than 1 years for chiary II and chiary III

6.1 Tables causes and frequency of final diagnosis

Table 5.1 Final Diagnosis of MRIs

		Frequency	Percent	Valid Percent	Cumulative Percent
Valid	Post traumatic	4	8.0	8.0	8.0
	Other	5	10.0	10.0	18.0
	Chiari I	14	28.0	28.0	46.0
	Chiari II	4	8.0	8.0	54.0
	Chiari III	3	6.0	6.0	60.0
	Spinal cord tumor	11	22.0	22.0	82.0
	Idiopathic Syringomyelia	3	6.0	6.0	88.0
	Kyphosis	1	2.0	2.0	90.0
	Kypho - scoliosis	3	6.0	6.0	96.0
	Multiple Diagnosis	2	4.0	4.0	100.0
	Total	50	100.0	100.0	

6.1 Table age with final diagnosis on MRI

Multiple diagnosis=kyphoscoliosis with diastomatomeia and CHF and one had kyphoscoliosis with tethered cord and pneumonia

Others= two patients had diastomatomeia and two tethered cord and one had post infectious

Table 6.2 Age * Final Diagnosis of MRIs Crosstabulation

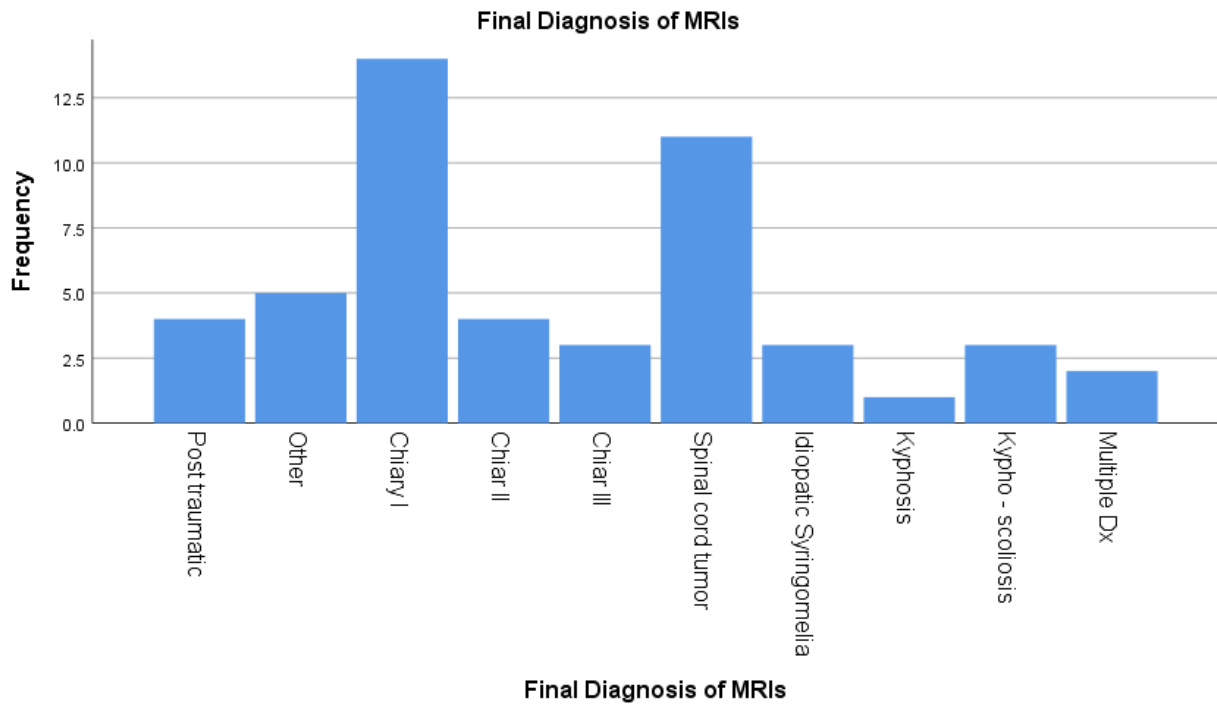
	Count	Total
Final Diagnosis of MRIs		

Age	Post traumatic	Other	Chiari I	Chiari II	Chiari III	Spinal cord tumor	Idiopathic Syringomelia	Kyphosis	Kypho - scoliosis	Multiple Dx	
< 1	1	1	0	2	1	1	0	1	0	0	7
1 - 10	1	2	1	2	2	1	0	0	1	2	12
11 - 20	0	0	1	0	0	5	1	0	0	0	7
21 - 30	0	0	2	0	0	3	0	0	2	0	7
31 - 40	1	0	7	0	0	0	1	0	0	0	9
41 - 50	0	2	2	0	0	0	0	0	0	0	4
> 51	1	0	1	0	0	1	1	0	0	0	4
Total	4	5	14	4	3	11	3	1	3	2	50

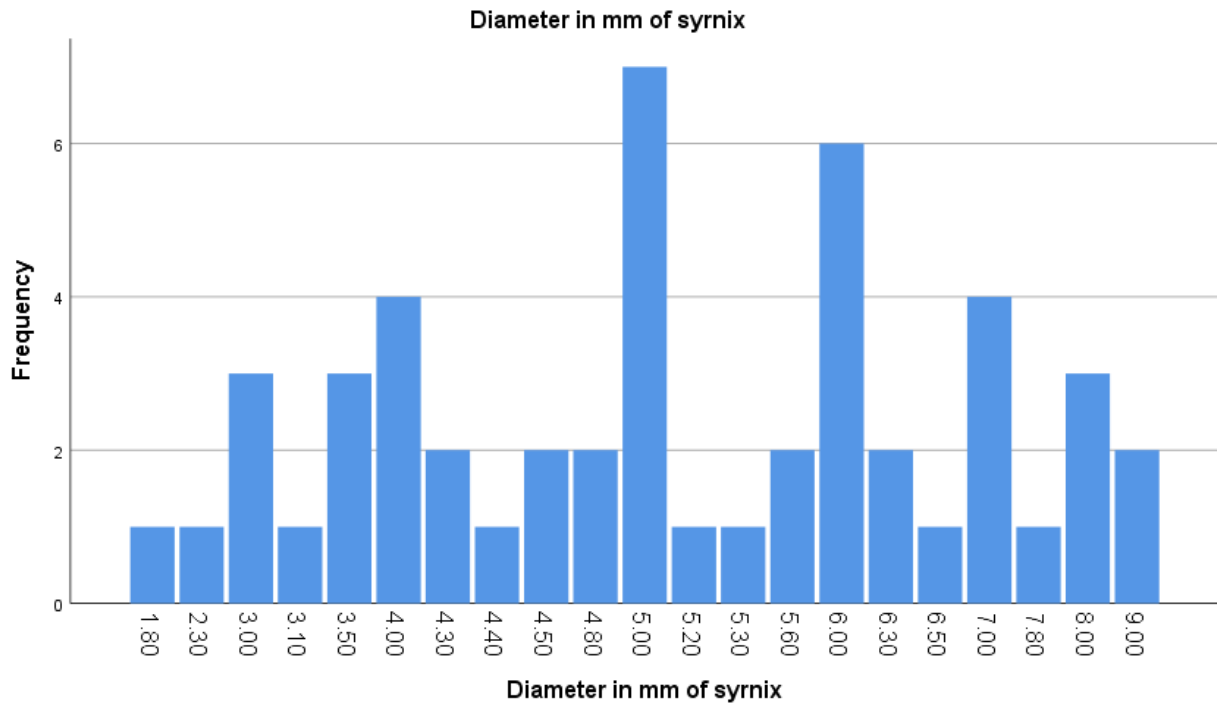
Multiple diagnosis=kyphoscolosiswith diastomatomeia and CHF and one had kyphoscolosis with tethered cord and pneumonia

Others= two patients had diastomatomeia and two tethered cord and one had post infectious

6.3 pie chart



Multiple diagnosis=kyphoscolosis with diastomatomeia and CHF and one had kyphoscolosis with tethered cord and pneumonia
Others= two patients had diastomatomeia and two tethered cord and one had post infectious



7Discussions

. Spinal syringes may result from several possible etiologies or, in idiopathic cases, may have no discernable etiology at all.[.71,72] It is well established that CM-I may lead to syrinx formation.[64,66,69,71,75,78], Most contemporary reports have suggested that abnormal flow of CSF at the craniocervical junction leads to syrinx formation.[66,71,75,77,78,80] However, given the relatively common incidence of CM-I and syrinx found in patients undergoing MRI, it is also possible that in some cases, a patient with low tonsil position also will have an intramedullary spine cyst for reasons unrelated to the CM-I. This idea is reinforced by clinical experience, in that syringes are quite variable in terms of size and position within the spinal canal. Since syringes may be caused by multiple associated conditions or even, for idiopathic syringes, have no known cause at all, it follows that not all syringes found in patients with CM-I are necessarily caused by the CM-I. There are several facts that establish the relationship between CM-I and syringomyelia.

our study are similar syringomyelia associated findings in Michigan university which were held on 2014 EC over interval of 11 years. A total of 271 patients with syrinx were identified the most common associated condition was 117 patients in [43.2%] followed by spinal dysraphism (20[7.4%]), tumor (15[5.5%]) and tethered cord (13[4.8%]). Eighty three patients (30.6%) did not have associated condition of brain or spinal cord their syringes were considered idiopathic First, syringes occur with significantly increased frequency in those with CM-I and reliably improves following CM-I decompression. 13,28,30 Many groups have plausibly described how changes in CSF flow at the foramen magnum can lead to syrinx formation.[10,20]

We have shown that CM-I-associated syringes are also usually distinct in location and size We found that CM-I-associated syringes had a larger mean axial dimension compared with other types and were more likely to have a cranial extent in the cervical spine. These findings are consistent with prior reports of CM-I-associated syringes.[10,28] Taken together, these facts imply that the mechanism of syrinx formation in CM-I-associated cases is likely to be different than in

syringes of a different morphological appearance. Mean syrinx length, however, was not useful in differentiating CM-I-associated syringes from other associated cause Spinal cord tumors maybe partly cystic.

The cystic and solid parts of tumors are seen to have different signal intensities; the locations and configurations of the cord expansion may also be eccentric and irregular.

In cases of syrinx associated with a spinal cord tumor the cyst had an appearance similar to simple hydromyelia, but the tumor itself often had abnormal signal on images.

In general, idiopathic syringes have a different morphology and location, tending to be narrower than CM-associated syringes and not as frequently located in the cervical spine. [72,73,74,76]

They are usually asymptomatic, incidental findings, and in most cases require no treatment. [72,73,74,76,] All patients in this series had a syrinx width of at least 1.8 mm. which is similar studied in the [72,73,74,76,]

There is no consensus definition for distinguishing a narrow syrinx from the very common finding of a dilated central canal on imaging. Some groups have used a minimum width threshold of 2 mm for an imaging diagnosis of syrinx; we used 1.8 mm width as the inclusion criterion for syrinx in this analysis [74,] We prefer the 1.8mm definition, since it is more likely to exclude very thin central spinal fluid collections that blend into the category of dilated central canal.

The threshold of 1.8mm allows for a greater focus on MRI findings that most would consider diagnostic of a syrinx.

Our study have similar findings with Conditions leading to syringomyelia can develop within the hindbrain or elsewhere [68,70]. The most common hindbrain lesion that results in syringomyelia is type I Chiari malformation, which develops within the foramen magnum [71]

8limitation of study

Poor quality of images and reports like demographic data

Artfactual image quality

Some image of a patient were missed and study depend on reports of image

7 conclusion

Syrinx location within the spinal canal also differed according to associated condition. Syringes associated with CM-I, CM-II, or CM III had a more superior cranial extent, usually in the cervical spine. Syringes associated with tethered cord and spinal dysraphism were more likely to have their cranial and caudal extents located more caudally.

The incidence of kyphosis/scoliosis did not differ significantly by associated condition, with the exception that patients with Chiari malformation were less likely to have scoliosis.

8 Recommendation

- This study gives initial insights for further study.
- This study: MRI is the modality of choice to see associated lesions.

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ANNEXES

ADDIS ABEBA UNIVERSITY CHOLEGE OF HEALTH SCIENCES
SCHOOL OF MEDICINE DEPARTMENT OF RADIOLOGY
A QUESTIONNAIRE ON MRI FINDING OF SYRNIX AND ASSOCIATED LESION

I. Demographic data 1. ID_____ 2 Age_____ 3 Sex

II Indication for MRI If possible symptoms of patient

- 1, Weakness 2.Back swelling 3.Gibbous Deformity4.Trauma to back or fall down accident
5 previous history of meningitides or TB Spondilities 6.Family members with
spinal MRI who has syrnix 7.Recommended from other imaging

III MRI findings

A On client MR image site of syrnix

- 1 Pones 2.Midbrain 3.Cranio-cervical junction 4. Cervical spinal cord 5 Cervicothoracic
upper thoracic spinal cord 6. Thoracic spinal cord 7.Lumbar spinal cord and conus-
medularies8.Whole spinal cord 9. Spinal cord ,pones and midbrain

B Measure width of syrnix on millimetermm

C spinal vertebral bone level from cranial to caudal

D The syrnix on spinal cord

1. Central midline 2. Off midline continues to center 3. Off midline only

E , Diameter of spinal cord on the site of syrnix relative to normal part

1. Expanded [increased] mention on centimeter 2. Narrowed [decreased] 3. Normal

F If any spinal canal lesion on area of syrnix, superior or inferior to the site of syrnix which is

a Extra-Dural 1. Mass if the diagnosis is known mention 2. vertebral bony lesion

3.others specifies

b Intra -Dural extra-medullary 1 Mass if the diagnosis is known mention 2

vertebral bony lesion 3 others specifies

G ,If there is vertebral bony lesion on x ray or spinal CT/MRI is known mention it

- 1 Lytic or sclerotic 2 Kypho-scoliosis 3 Spondilosis 4 Spondilosis with spondilolistesis

5 Bony defect 6 If there is diagnosis specify

H, If any associated spinal cord lesion

1. Tethered cord 2 .Diastematomelia type I 3.Diastematomelia type II 4.Mass
5.Post--surgical change 6. Infectious

I If there is brain MRI cerebellum tonsil down ward below foramen magnum

1. Greater than 5mm mention 2.less-than 5mm

J cerebral ventricular dilation and associated findings

a, Hydrocephalus with cerebella tonsil herniation 1 Yes 2 No

b, a ,with posterior fossa cystic lesion 1 Yes 2 No

c, a, with posterior fossa mass 1 Yes 2 NO

K,Final of diagnosis of MRI 1, Chiary malformation writes one a ,Chiary I b, Chiary II
c, Chary III d Chiary IV

L .Final of diagnosis of MRI

1. Post traumatic, 2. Spinal canal mass tumor specify likely diagnosis

3. Spinal cord tumor specify likely diagnosis. 4. Post infectious specify if known

5. Familial syrinx, 6.Idiopathic syringomelia 7.Kyphosiss 8.Kyphoscolosis

9. If multiple write numbers here _____

_____10. Others specify