

# Concepts in the Management of Syringomyelia

<sup>1</sup>Abhidha Shah, <sup>2</sup>Abhinandan Patil, <sup>3</sup>Shashi Ranjan, <sup>4</sup>Atul Goel

## ABSTRACT

**Objective:** The authors analyze their experience with syringomyelia. The treatment was focused on identification of the primary etiological factor and its treatment.

**Methods:** Depending on the etiological factors and treatment considerations the series was classified into three groups. Group 1 had cases where there was no definite demonstrable etiological factor. Group 2 cases had basilar invagination and/or Chiari malformation, and Group 3 consisted of cases where the syrinx was secondary to an obvious aetiology, such as a mass lesion either in the posterior cranial fossa or in the spine or a severe kyphotic spinal deformity. Post-traumatic syringomyelia and syrinx in association with spina bifida were not studied. There is a significant subgroup where no cause is identified when evaluated by conventional radiological parameters. However, atlantoaxial dislocation was identified when assessed by Goel classification of facet alignment.

**Results:** In general, in Group 1 (or in idiopathic group), atlantoaxial instability was identified and was accordingly treated. In Group II, atlantoaxial instability was considered to be defining phenomenon. Accordingly atlantoaxial fixation was the treatment. In Group III the treatment was focused on the etiological factor. It was identified that direct syrinx drainage was not only not useful, but was harmful. It was observed that clinical outcome rather than radiological improvement is the reliable indicator of the surgical result.

**Conclusion:** Syringomyelia is 'never' a primary pathological event but is secondary to a known or unknown (or unidentified) pathology. Treatment of the primary etiology is the goal in management of this condition

**Keywords:** Atlantoaxial instability, Basilar invagination, Chiari formation, Syringomyelia.

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## INTRODUCTION

The subject of syringomyelia has been known, evaluated and extensively discussed for over a century. A number of

clinical articles and reviews on the subject are a testimony to the complexities of the issues involved. The investigations for confirming the presence of syringomyelia have evolved over the period. More than anything else, the treatment for the entity of syringomyelia varies with each individual surgeon. The very fact that no consensus regarding the best treatment modality that can be most suitable and uniformly applicable has yet been identified points to the fact that the pathogenesis of syringomyelia has not yet been entirely or clearly understood.

## Historical Origin

The word syrinx is a Latin word derived from the Greek word *surrinx*, which means a series of panpipes. The origin of the word syrinx is in classical Greek mythology. According to this syrinx was a nymph who was pursued by the ardent God Pan. To maintain her chastity she ran to a river's edge and asked for help from the river nymphs. The nymphs transformed her into hollow water reeds. The reeds made an eerie sound when the God's breath blew across them. Pan cut the reeds to make the first set of pan pipes which henceforth came to be known as "syrinx".

## Origin in medical literature

In 1827, Charles-Prosper Ollivier d'Angers coined the term "syringomyelia" to describe the presence of a pathological cavity in the spinal cord.<sup>1</sup>

## THEORIES OF SYRINX FORMATION

The origin of syrinx fluid has been a matter of speculation from its inception. Several authors have proposed various theories to explain the source and propagation of the syrinx fluid.

## Gardner's Hydrodynamic Theory

In 1965, Gardner published the hydrodynamic theory to explain the pathophysiology of syringomyelia.<sup>2</sup> He postulated that it was not the mean increased pressure in the ventricles that caused the syrinx formation but rather the water hammer effect of the sudden spurt of ventricular fluid imparted to it by each pulse beat of the choroid plexus. The syrinx develops by a method of hydrodissection similar to that which is responsible for the development of the subarachnoid space in the embryo.

<sup>1</sup>Assistant Professor, <sup>2,3</sup>Senior Resident, <sup>4</sup>Professor and Head

<sup>1-4</sup>Department of Neurosurgery, Seth G.S. Medical College and K.E.M Hospital, Parel, Mumbai, Maharashtra, India

**Corresponding Author:** Abhidha Shah, Assistant Professor, Department of Neurosurgery, Seth G.S. Medical College and K.E.M Hospital, Parel, Mumbai, Maharashtra, India e-mail: abhidha@gmail.com

## William–Theory of Craniospinal Pressure Dissociation

According to the craniospinal dissociation theory, during moments of raised intra-abdominal pressure there is venous congestion of the epidural veins causing engorgement around the dural sac.<sup>3,4</sup> This in turn causes displacement of the CSF upwards towards the cranial cavity. In normal individuals this excess CSF would just as easily flow downwards after the episode causing raised intra-abdominal pressure has ended. In patients with outflow obstruction, the site of obstruction acts as a valve not allowing the CSF to flow back. Thus a pressure differential is created causing a suck effect. This suck effect causes the CSF from the fourth ventricle to flow down through the obex into the central canal thus causing syringomyelia. Once the fluid is present within the cord cavity and it reaches a critical size, it pulsates both upwards and downwards, in response to fluid movements and pressure changes in the subarachnoid space, the most influential being the venous pressure changes. This movement “slosh” causes both upward and downward propagation of the syringomyelia.

However for these theories to be true, for syringomyelia to occur there would have to be a patent communication between the fourth ventricle and the central canal which has not been seen frequently. It has been argued by some that the communication may have been present initially but closes off after a period of time.

## Ball and Dayan’s Theory

Ball and Dayan proposed that due to an outflow block there is an obstruction in the rostral flow of spinal CSF from the spinal subarachnoid spaces to the intracranial subarachnoid space during periods of raised intrathoracic pressure.<sup>5</sup> Due to this there is an increase in the spinal CSF pressure which pushes the spinal CSF along extracellular pathways into the spinal cord surface thus initiating and propagating the syringomyelia.

## Piston Theory

In 1994, Oldfield proposed another theory to explain the formation and propagation of the syringomyelia based on the normal cardiac cycle.<sup>6</sup> In normal individuals, during systole the brain expands after receiving blood. This causes the CSF to flow from the 4th ventricle into the cisterna magna and then onwards into the upper cervical canal through the foramen magnum. During diastole the CSF flows upwards from the spinal canal to the cranial cavity across the foramen magnum. When there is obstruction at the region of the foramen magnum there is impedance to this rapid to and fro movement of CSF in the subarachnoid space across the foramen magnum during systole and diastole. During systole the brain expansion is

accommodated by an abrupt piston like caudal movement of the tonsils. As there is occlusion of the rapid upward movement of the CSF there is a partially isolated spinal subarachnoid space. This movement of the tonsils imparts a pressure wave to the spinal subarachnoid space causing fluid movement of CSF into the cord and also propelling the syrinx fluid in the cord inferiorly.

## Intramedullary Pulse Pressure Theory

According to this theory the driving force of syringomyelia is the systolic CSF pulse pressure, which is the pressure wave of CSF displaced from the head during arterial pulsations.<sup>7</sup> As there is obstruction of the subarachnoid space, there is a significant decrease in pressure transmission to the distal CSF spaces. This in turn increases the systolic CSF pulse pressure, which gets transmitted to the spinal cord tissue in close proximity to the obstruction. This increased pressure in the spinal cord and the decreased pressure in the nearby CSF space distends the spinal cord just below the site of block. Also the increased systolic CSF pressure is simultaneously transmitted to the spinal cord at the level of the obstruction thus distending the cord above the level of obstruction.

## Pathogenesis

Syringomyelia is a relentlessly and progressively growing intramedullary phenomenon that is related to chronic or longstanding pathological alterations in and around the spinal cord.<sup>8,9</sup> Factors like trauma, infections, tumors and craniovertebral junctional abnormalities have been incriminated to be causative. Chiari formation and basilar invagination are frequent neural and musculoskeletal accompaniments of syringomyelia. We recently identified that musculoskeletal events of the craniovertebral junction and spine that includes basilar invagination, short neck and torticollis and neural events like Chiari formation and syringomyelia are not primary issues but are secondary and natural protective processes to one single pathological event of atlantoaxial instability.<sup>8,10,11</sup>

There is a significant subgroup where no cause is identified and syringomyelia in such patients is labeled to be “idiopathic.”<sup>12-16</sup>

## Natural Protection

In the year 2000, Goel identified syringomyelia as a natural protective process and ‘always’ a secondary issue to a known or an unknown (or unidentified) primary problem.<sup>8,9</sup> A number of primary etiological factors have been known to be associated with syringomyelia. Essentially, he recognised that syringomyelia is not a pathological or adverse event, but a protective self neural destruction that works in the interest of the human body and an attempt by nature to stall or delay the effects

of the primary pathological process. The 'self-neural destruction' or syringomyelia may not even be recognised clinically for a period of time or patients with huge syrinx cavity may present with only marginal clinical symptoms, as it seems that relatively 'less important' neural pathways are sacrificed in favour of the more important ones.

## CLINICAL PRESENTATION

Pain in the neck, shoulders and hand are the more prominent and constant presenting clinical features. Wasting and weakness of hands and spasticity and weakness in the legs can be present in more longstanding cases. Classically described sensory loss that includes affection of posterior column sensations and affection of the temperature and pain sensations in the shoulders and hands are more characteristic presenting symptoms. Despite the presence of syringomyelia and reduction in the neural girth, the symptoms are remarkably less and few.

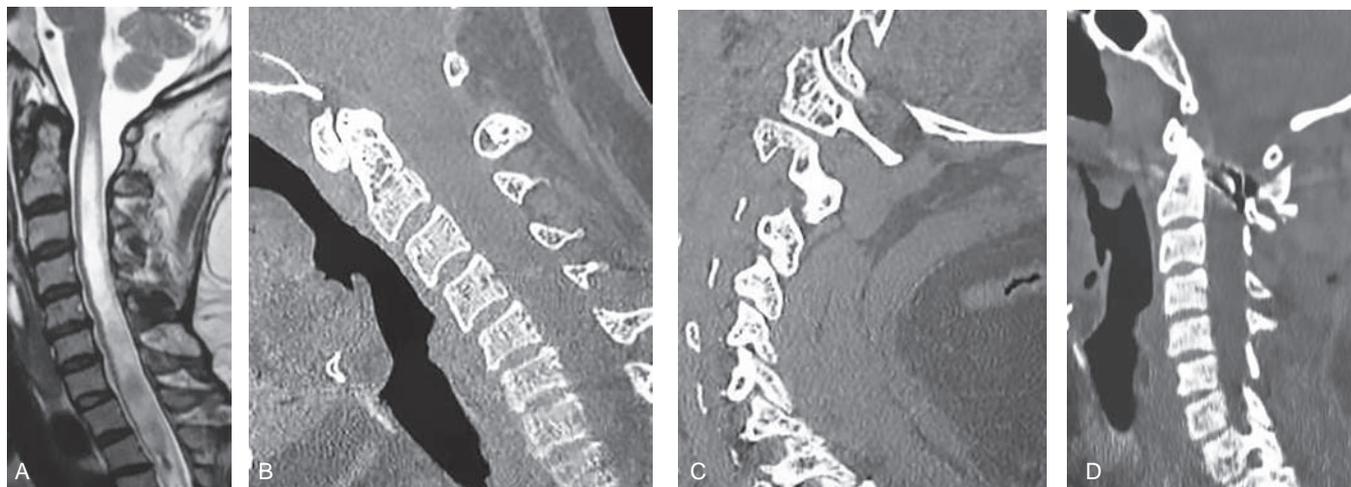
## Classification

In the year 2000, Goel classified syringomyelia into three groups depending on the identified etiological factor.<sup>8</sup> Group 1 cases included those where no definite cause could be identified. This formed a relatively small but a distinct subgroup of patients. Group II cases had basilar invagination and/or Chiari malformation; and Group III consisted of cases where the syrinx was secondary to an obvious etiology, such as a mass lesion either in the posterior cranial fossa or in the spine or a severe kyphotic spinal deformity. Post-traumatic syringomyelia and syrinx in association with spina bifida were not studied.

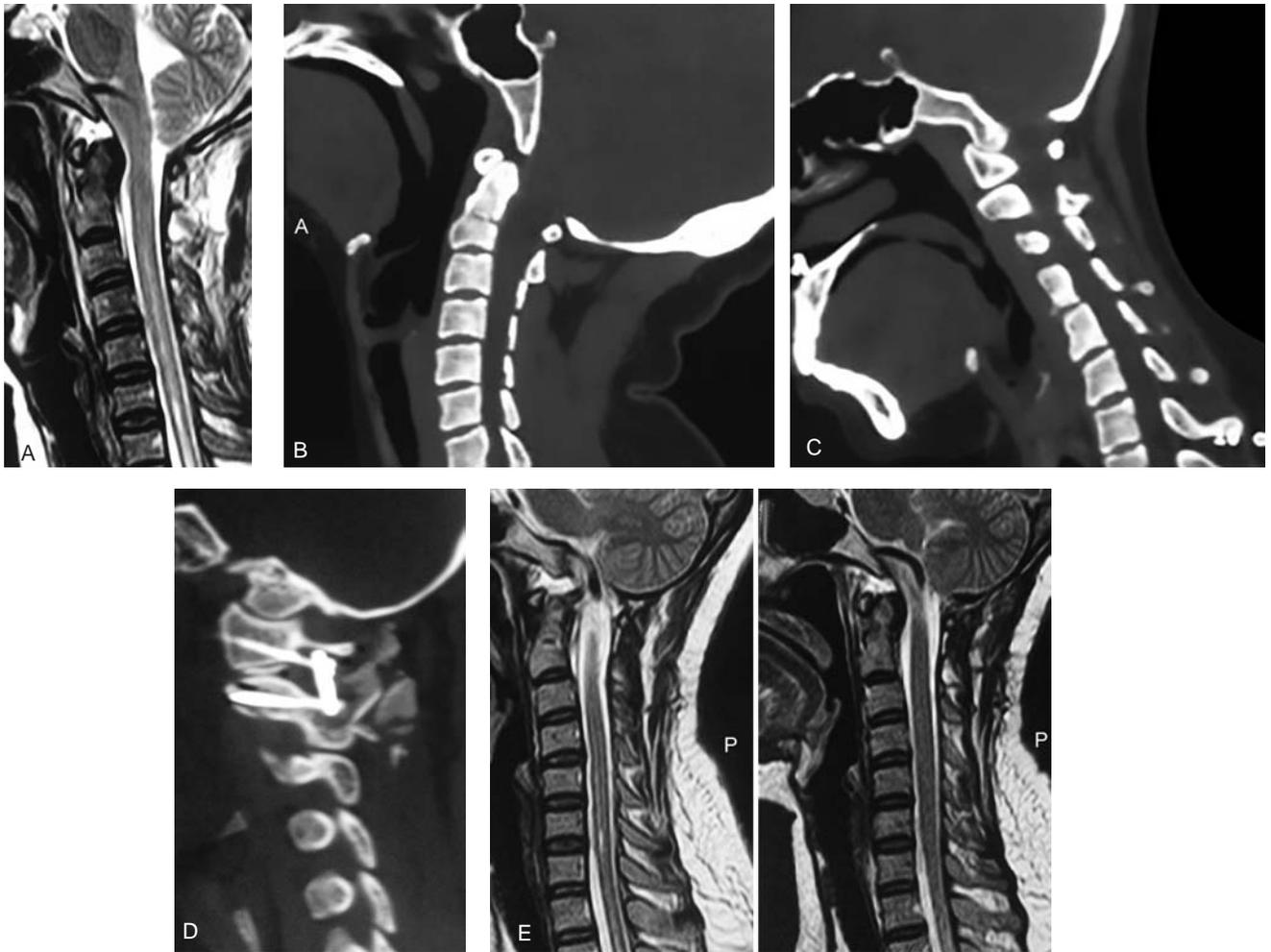
## Group 1—Idiopathic Syringomyelia (Figs 1 and 2)

There are several articles in the literature that discuss "idiopathic" syrinx.<sup>12-16</sup> Alteration in the cerebrospinal fluid flow dynamics in the region of craniocervical junction related to chronic inflammation is the most frequently speculated cause of such syringomyelia. As no definite cause is identified, the treatment recommended is usually shunt surgery that involves drainage of the syrinx cavity. There have been reports where opening of the foramen magnum and lysing the arachnoidal adhesions have been recommended. However, no universally accepted treatment protocol is yet available.

We recently reported a series of nine cases having 'idiopathic' syringomyelia and assessment by conventional radiological or clinical parameters did not reveal direct evidence of any incriminating causative factor.<sup>17</sup> We observed that there was atlantoaxial instability in these cases and atlantoaxial fixation resulted in remarkable clinical improvement. The commonly recognized radiological parameter of identification of atlantoaxial instability is to observe abnormal alterations in atlantodental interval and to identify indentation of the odontoid process into the cervico-medullary cord. We observed that atlantoaxial instability can be identified by focusing the attention on the facets. We discussed three types of atlantoaxial facetal dislocation.<sup>18,19</sup> Type A atlantoaxial facetal dislocation is the more common form of dislocation wherein the facet of atlas is dislocated anterior to the facet of axis. We identified such a form of dislocation as atlantoaxial facetal listhesis and related it to more frequently identified and recognized entity of lumbosacral spondylolisthesis.<sup>20-22</sup> In this form of instability, the atlantodental interval increases and the odontoid process indents into the neural structures. Such disloca-



**Figs 1A to D:** Images of a 49-year-old female patient (A) T2-weighted MRI showing extensive syringomyelia. There is no craniocervical bone or soft tissue abnormality; (B) Sagittal image of CT scan, with the head in flexion position, showing no evidence of atlantoaxial instability when assessed by parameter of atlantodental interval abnormality; (C) Sagittal CT with the cut passing through the facets showing type B facetal instability; (D) Postoperative CT scan after atlantoaxial fixation



**Figs 2A to E:** Images of a 37-year-old female patient (A) T2-weighted MRI shows normal bone and neural architecture in the region of foramen magnum and craniocervical junction. Syringomyelia can be noted; (B) CT scan shows no evidence of instability as measured by atlantodental interval alteration; (C) CT scan of the head in flexed position showing Type B facetal dislocation; (D) Postoperative CT scan showing the implant and fixation; (E) Delayed postoperative T2-weighted MRI showing reduction in the size of the syrinx

tion is associated with relatively acute clinical presentation. Type B atlantoaxial facet dislocation is when the facet of atlas is dislocated posterior to the facet of axis. Type C atlantoaxial facet dislocation is when the facets are in alignment but the atlantoaxial joint is unstable. Such instability is also called axial or central instability. Instability in Types B and C is identified during direct observation and manipulations of the facets during surgery that involves fixation of the lateral masses. As in both Types B and C, the odontoid process is not displaced posteriorly or atlantodental interval is not abnormally increased and there is no direct indentation of the neural structures, they are associated with more chronic form of musculoskeletal and neural alterations.

We recently evaluated atlantoaxial instability in chronically “unstable” clinical situations like basilar invagination, wherein there is a longstanding alteration in the physical musculoskeletal and neural structural form.<sup>10</sup> We analyzed the role of atlantoaxial instability in pathogenesis of cases having Chiari malformation with or without the association of syringomyelia and with or

without the association of bone abnormality in the craniocervical junction.<sup>11</sup> On the basis of our observation it was concluded that Chiari malformation may have “air-bag” type of naturally protective effect on the spinal cord and may have a role in preventing pinching of critical neural structures between bones.<sup>23</sup> We also speculated that syringomyelia may be a natural self-neural destruction that ultimately works in the best interest of the patient.<sup>8</sup>

In our presented series, we identified that eight cases had Type B and one patient had Type C atlantoaxial facet dislocation. There was no or only “insignificant” evidence of cord compression at the level of foramen magnum. Our experience of manual handling the facets of atlas and axis and the atlantoaxial joint for over 30 years and surgically treating over 2000 cases having instability of the region, we identified that the joint in these cases was clearly unstable.<sup>20,21,24,25</sup> Moreover, fixation of the joint resulted in an immediate postoperative relief from symptoms. Remarkable reduction in the size of the syrinx cavity in most cases without any dural violation

or syrinx cavity manipulation does suggest the validity of the observation.

Essentially, the observation leads to the suggestion that atlantoaxial instability can be a possible cause in cases having “idiopathic” syringomyelia. The reason as to why Chiari malformation is not associated in such cases is unclear. Our observations suggest that investigations should focus the attention on the status of alignment of facets of atlas and axis. Whilst some degree of facet mal-alignment can be within the range of normalcy, its presence in association with syringomyelia can have clinical and therapeutic significance. In the presence of even subtle evidence of bone abnormalities in the craniovertebral junction, like bone fusions or malformations, even normal facetal alignment can signify instability. We labeled such form of facetal instability as axial or central instability (Type C instability).

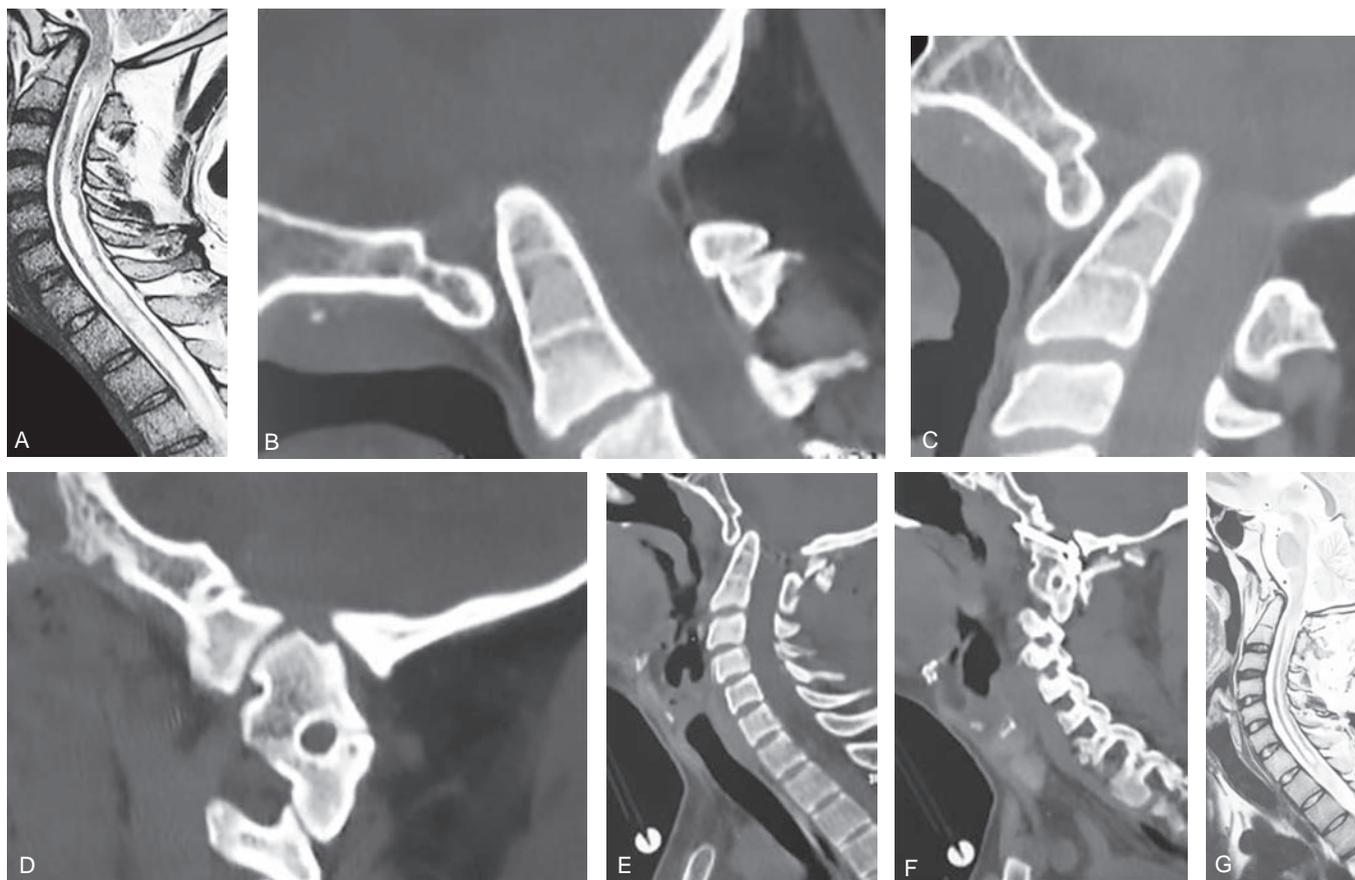
**Group 2–Syringomyelia in association with Basilar Invagination and Chiari formation (Fig. 3)**

This group forms a major subgroup of patients with syringomyelia. In these cases it essentially appears that syringomyelia is a tertiary event to the primary basilar invagination and secondary Chiari malformation. The

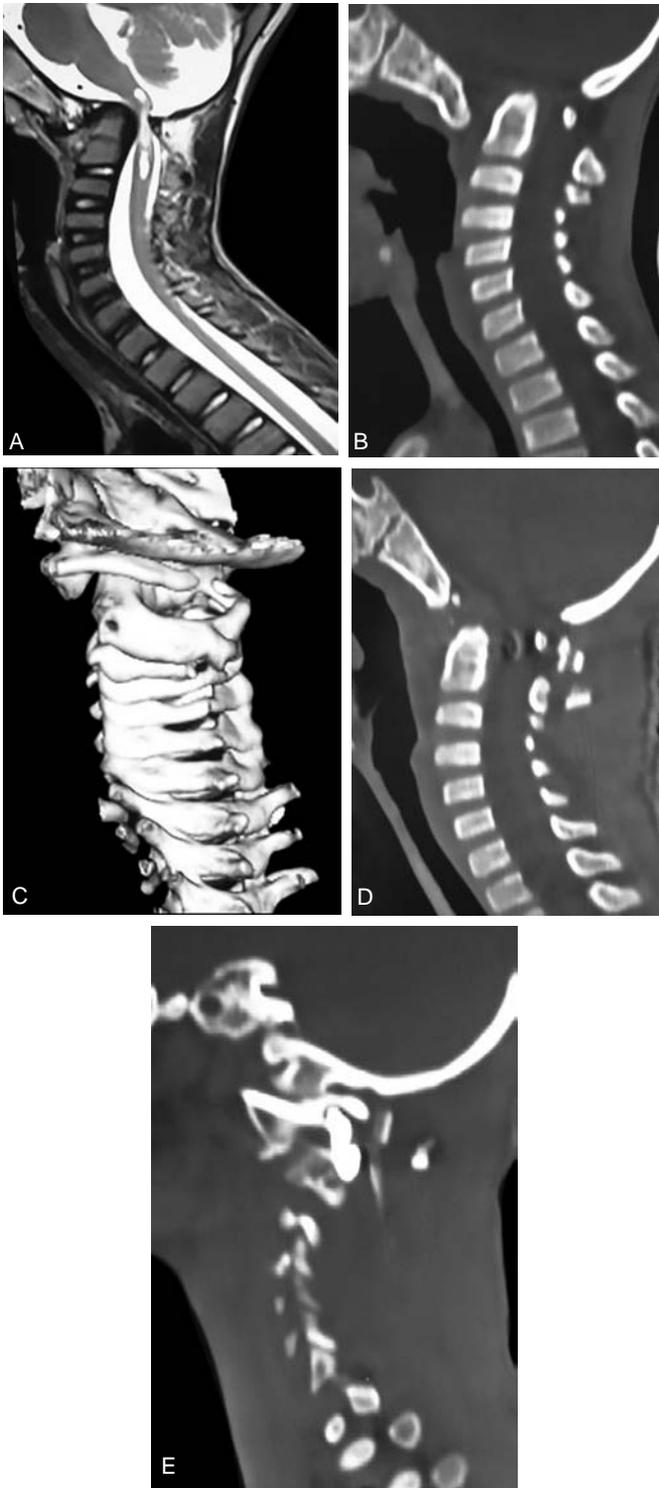
treatment modality in these cases is craniovertebral stabilization.

**External Syringomyelia: Introduction of a new term (Fig. 4)**

We have identified that there are several alterations in the spine and the posterior cranial fossa in conjunction with basilar invagination and Chiari formation.<sup>26-28</sup> The spinal canal increases in its circumferential dimensions and the spinal cord reduces in its girth. Similarly, the clivus becomes short resulting in the short head. Like the neck that becomes short and spinal canal that becomes larger, the posterior cranial fossa becomes shorter in its vertical height but longer in its transverse and anteroposterior dimensions. In addition, the neural structures in the posterior fossa that include cerebellum and brainstem become thin and atrophic. The overall effect increases the space available in the spinal canal and posterior cranial fossa that is occupied by CSF. In the spinal canal, the increased collection of CSF may be in the extramedullary space (external syrinx), inside the spinal cord (syringomyelia) or both inside and outside the spinal cord.<sup>26-28</sup> An excessive amount of CSF is also present in the posterior



**Figs 3A to G:** Images of a 19 year old male patient (A) T2 weighted MRI showing basilar invagination cord compression and syringomyelia; (B) CT scan with the head in flexion showing basilar invagination with assimilation of atlas; (C) CT scan with the head in extension showing no reduction in the basilar invagination; (D) CT scan with cut passing through the facets showing Type A facetal instability; (E) Post-operative CT scan showing the craniovertebral re-alignment; (F) Post-operative CT scan showing the implant; (G) Post-operative MRI showing reduction in the size of the syrinx



**Figs 4A to E:** Images of a 7 year old male child (A) T2 weighted MRI showing Group A2 basilar invagination and cord compression. External syringomyelia and external syringobulbia can be observed; (B) CT scan showing basilar invagination; (C) 3D CT scan showing type I atlantoaxial facet instability. Bifid posterior arch of atlas can be observed; (D) Postoperative CT scan showing reduction of basilar invagination and fixation; (E) Postoperative CT scan with the cut passing through the facets showing the implant.

cranial fossa, inside the brainstem (syringobulbia), anterior and around the brainstem (external syringobulbia) and around the cerebellum.<sup>26-28</sup> The cerebellum, more in its superior vermis and superior cerebellum, becomes

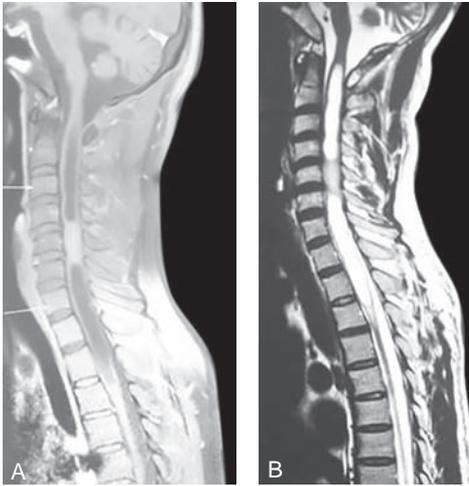
atrophic allowing an increased collection of CSF. Superior vermis is atrophied, but the herniated part of the tonsil is never atrophied, probably because a firm cerebellar tissue is necessary to provide a compact cushion.

Atrophy of the neural structures at the point of compression by the odontoid process and in the spinal cord and brainstem seem to be a natural response to reduce the girth of neural structures that allows it to accommodate the intruding odontoid process and make the neural tissues resilient and stronger. Essentially, we speculated that the bony spinal canal and posterior fossa decrease in vertical height and neural structures (spinal cord, brainstem, and cerebellum) become thinner and longer in length, allowing the neural structures to course over the odontoid process in a relatively stretch free traverse. The direction of the angulation of the odontoid process influences the presence of CSF within or outside the neural tissues.<sup>27</sup> We observed that more severe is the angulation of the odontoid process and more severe is the compression of the neural structures more is the incidence of the presence of external syrinx. Essentially, it seems that external syrinx and excessive collection of CSF in the posterior cranial fossa allows the neural structures to float away from the compressing odontoid process. Reduction of girth of the neural structures seems to be a result of escape of CSF in the central spinal canal and fluid inside and outside the neural fibers in the spinal cord. In this respect, it seems that the pathogenesis and function of CSF, whether it is inside the cord in the form of syringomyelia (or syringobulbia) or whether it is outside the spinal cord in the form of the external syrinx (or external syringobulbia) is essentially the same.<sup>26-28</sup> Syringomyelia is more often seen when the odontoid process is either in its normal position or is vertically herniated into the posterior cranial fossa. External syringomyelia is more often when the odontoid process is posteriorly angulated or horizontal in its lay. The marked resilience of the neural structures and preservation of the neural function despite the severe reduction of neural dimensions are unique protective ways of nature. Our observations suggest that atlantoaxial dislocation is the only craniovertebral anomaly. All other bony and soft tissue alterations are secondary to atlantoaxial instability and are protective in their function.

### Group 3—Syringomyelia Secondary to an Obvious Etiology

#### *Tumors and Syringomyelia (Figs 5 and 6)*

Syringomyelia is frequently associated with intramedullary tumors. As syringomyelia is a chronic and a relent-



**Figs 5A to B:** Images of a 45 year old male patient (A) T1 weighted MRI showing the cervical intramedullary tumor and the syringomyelia; (B) T2 weighted MRI

lessly growing process, its presence is an indicator of chronicity of the entire process and slow growing or benign nature of the tumor. Syringomyelia is associated with intramedullary tumors like ependymomas or hemangioblastomas and only rarely with intramedullary astrocytomas. Presence of syringomyelia in cases with intramedullary tumor is a positive indicator of presence of benign tumor and the possibility of a positive long-term outcome. The relationship of the extent of syrinx with the further histological categorisation of the tumor is not yet entirely clear. The treatment in these cases, is removal of the tumor without manipulation of the syrinx.

### Scoliosis and Syringomyelia

Scoliosis can be a presenting clinical feature of syringomyelia. In such cases, it is sometimes difficult to identify if syringomyelia is the cause or the effect of scoliosis. Our current evaluation of the subject suggests that syringomyelia is the primary cause of scoliosis in most cases.

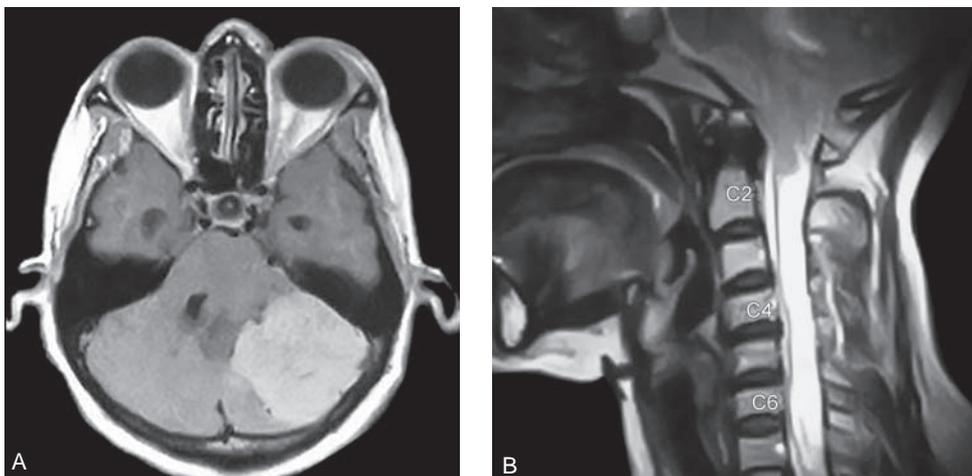
'Idiopathic' syringomyelia in such cases may be related to atlantoaxial instability. Treatment of atlantoaxial instability can result in reduction of scoliosis or can stall its progression.

### Special Surgical issues

Atlantoaxial fixation using the technique described by Goel and Laheri in 1994 provides a solid and stable ground for bone fusion.<sup>24,25</sup> However, atlantoaxial fixation surgery in cases having syringomyelia and external syringomyelia is relatively difficult and technically challenging. The spinal cord is lax and the extradural venous plexus is large. This can sometimes result in excessive bleeding during the exposure of the lateral masses for the surgical procedure. Careful subperiosteal dissection and avoidance of use of sharp cutting surgical tools can reduce venous bleeding. Packing of the extradural space with Surgicel and/or gelfoam can help in reducing the blood loss. Avoidance of pressure ventilation during surgery can also assist in reduction of bleeding. Head can be placed in higher position than in the usual cases where atlantoaxial fixation is done. The procedure essentially involves opening of the joint, denuding of the articular cartilage, packing of bone graft within the joint cavity and subsequent application of instrumentation.

### Clinical Outcome

Clinical outcome following atlantoaxial fixation is remarkably satisfying. In majority of properly selected cases, the clinical improvement can be observed in the immediate postoperative period after reversal from anesthesia. Pain in the hand and shoulders, tingling paraesthesiae in the limbs and limb weakness and spasticity can improve in the postoperative phase. The improvement is progressive.



**Figs 6A and B:** Images of a 39 year old female patient. (A) Axial contrast enhanced MRI showing the tentorial meningioma; (B) T2 weighted sagittal MRI of the patient showing the Chiari formation and syringomyelia

## Radiological Outcome

Despite the fact that clinical outcome can be dramatic, radiological demonstration of reduction in the size of the syrinx is not as predictable. The syrinx size reduces in approximately 30 percent of cases on postoperative imaging in about 30 percent cases. Our delayed imaging protocol now suggests that the syrinx size ultimately reduces in almost 80 to 90 percent cases when assessed radiologically at 6 months postoperative period.

## CONCLUSION

Syringomyelia is 'never' a primary pathological event but is secondary to a known or unknown (or unidentified) pathology. Treatment of the primary etiology is the goal in management of this condition. Atlantoaxial instability is a major cause of syringomyelia. Ignoring this fact can lead to wrong treatment and poor surgical outcome. On the contrary, atlantoaxial stabilization can result in an immediate postoperative recovery in neurological symptoms.

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