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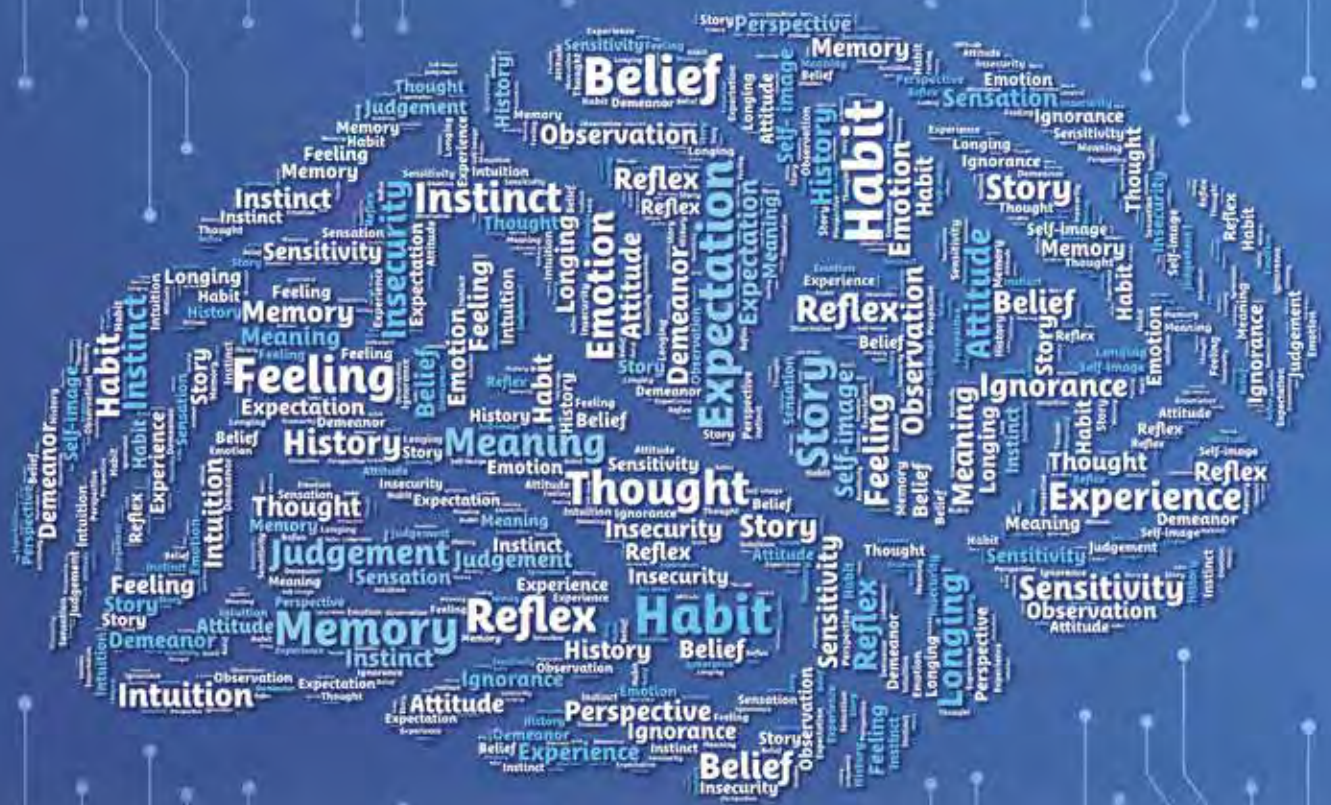
Fortis

# Clinical Connect

Fostering a culture of innovation and excellence

Neuro Sciences Special

## Exploring Advances in Neuro Sciences




## IN THIS ISSUE

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# Doctor's Day: MD & CEO's Message

Dear Colleagues,

I am delighted to extend heartiest Doctor's Day greetings to all our Clinicians.

On July 1 every year, we celebrate National Doctor's Day to thank and acknowledge the invaluable contribution of doctors in keeping the nation healthy.

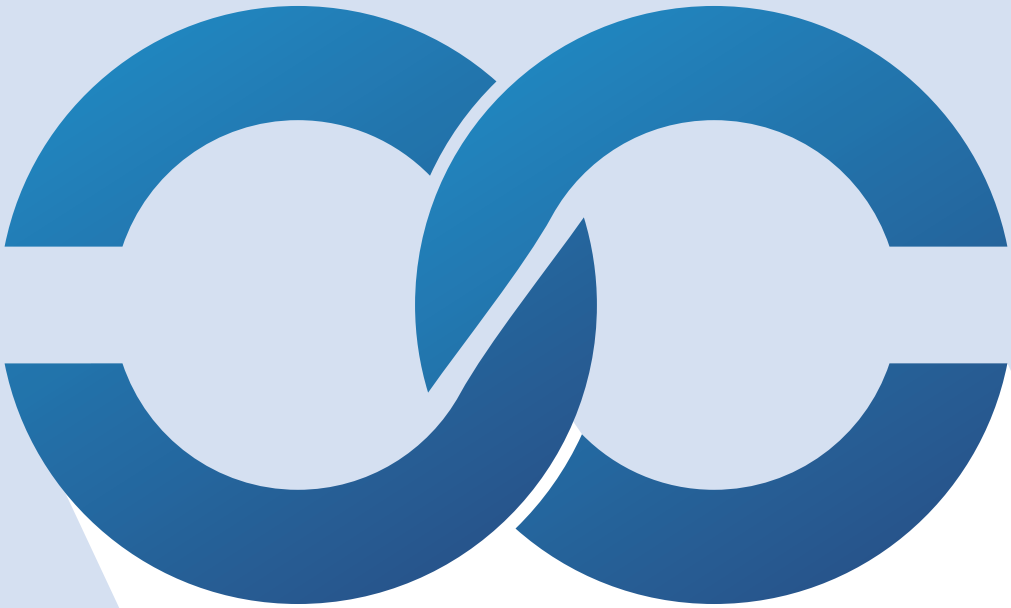
It is not easy to be a doctor and it is certainly not easy to be a good doctor. As we celebrate Doctor's Day, let us ponder for a moment on the qualities that make a doctor worthy of admiration. To my mind, the continuous quest for excellence, compassionate commitment towards the wellbeing of patients and a never-give-up attitude are some of the endearing hallmarks of a great doctor. We are fortunate to have talented teams across our network that live up to these values and are an inspiration in themselves.

I am proud of the path-breaking and innovative work our clinicians are doing every day. Thanks to all our doctors, we are today recognised for our clinical excellence. As an organisation, we are committed to invest in creating an enabling infrastructure that allows our doctors to do their best. We are rapidly adding new equipment, technology and clinical programmes at our hospitals, and are ramping up the infrastructure. We plan to make many more such investments in future.

I once again thank all our doctors for their incredible contribution in making our organisation one of the most preferred healthcare destination for patients from all over the world.

Happy Doctor's Day and best regards,

**Dr A. Raghuvanshi,**
MD & CEO
Fortis Healthcare Limited



# INSPIRATION

# Message

**Dr Praveen Gupta**
Principal Director & Head - Neurology
Fortis Memorial Research Institute,
Gurugram

On behalf of Fortis Medical Council, it is an honour and great pleasure to present 'Clinical Connect,' the bi-monthly newsletter. It is an endeavour to share the excellence of various esteemed clinicians of Fortis Healthcare.

We have been through very challenging times in healthcare due to COVID pandemic but with the fortitude and guidance of leadership team and diligence of clinicians. We have managed well. 'Clinical Connect' is a great attempt to pool the talents of nationally and internationally acclaimed clinicians and disseminated to Fortis health profession at large. It provides a unique opportunity to learn from each other.

The Fortis Neurology Council is dedicated to make neuroscience services uniformly state of art across Fortis Network Hospitals. It endeavours to create opportunities to share and refer patients, to facilitate their wellbeing. It also creates uniform protocols for better treatment across the network. It will enable to share the data to allow us publish research papers and learn from each other.

This newsletter spearhead by a powerful editorial team will be enriched by generous contributions from all esteemed clinicians. It will lay the foundation of Fortis Medical excellence.

# Message

**Dr Rajakumar Deshpande**
Director – Neurosurgery
Fortis Hospital, Bannerghatta Road,
Bangalore

**Fortis Neuroscience - Spearheading Outcomes**

**Dear Colleagues,**

In this issue of 'Clinical Connect,' the Fortis Neurosciences experts showcase their talent and might in managing a plethora of brain and spine disorders. This reflects the deep expertise that spans the many fields that encompass this difficult subject.

It is often said "a man travels the world over in search of what he needs and returns home to find it". The vast range of surgeries that Fortis neurosurgeons perform every other day is a testament to this. In this regard, it may be worthwhile to consider translating this enormous potential into something tangible for training post-graduate students who pass through our portals. It would be satisfactory, indeed, to impart this in a structured curriculum that is meaningful and compares with the very best institutes.

The entire range of subspecialty work that is performed by our neurosciences team cannot be published in one issue. A brief "peep" into the various nuances that make a successful outcome is being presented. Some of the cases are noteworthy for the complex nature which has been skilfully managed. For example, microsurgical techniques that requires years to hone to the highest level, are showcased when treating brain tumours. A similar masterly endeavour is seen in the cases of vascular disorders where timing is everything in the outcome.

Also, neurosciences the world over is changing rapidly. Technologies that are mind boggling are being introduced into daily practice. For example, robotics in neurosurgery, especially in advanced spine surgery, is no more a distant vision. Many institutes the world over, even in India, are using this invention to accurately place pedicle screws to treat complex spine deformities whose treatment were unthinkable even a few years ago. There is an urgent need to reflect on having such technologies in our institutions in the near future. It is imperative that we progress in giving the best to our patients.

It is a great pleasure to be a part of this scientific presentation of the Fortis Neurosciences.

# Message

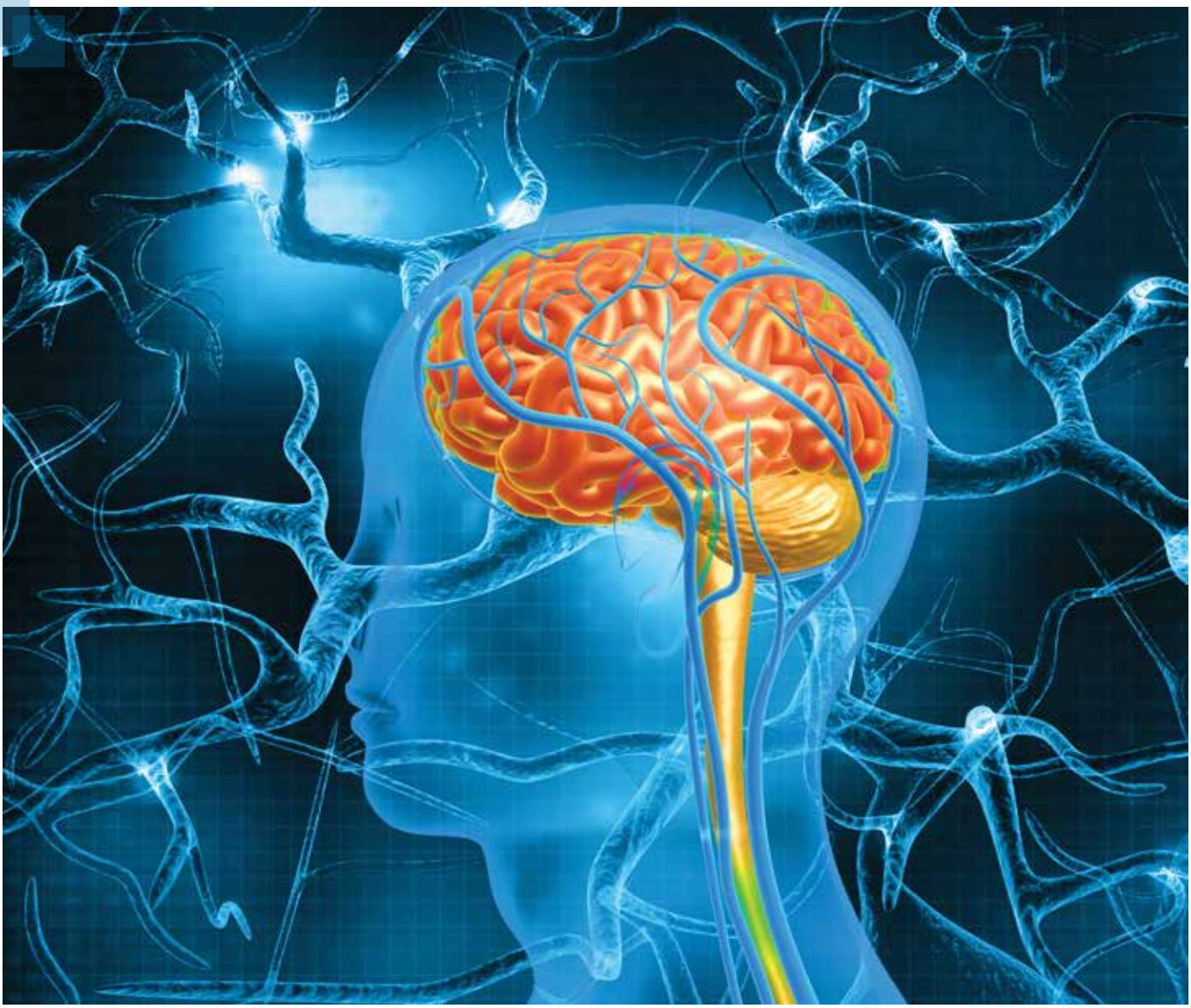
**Dr Sandeep Vaishya**
Executive Director &
Head - Neurosurgery
Fortis Memorial Research Institute,
Gurugram

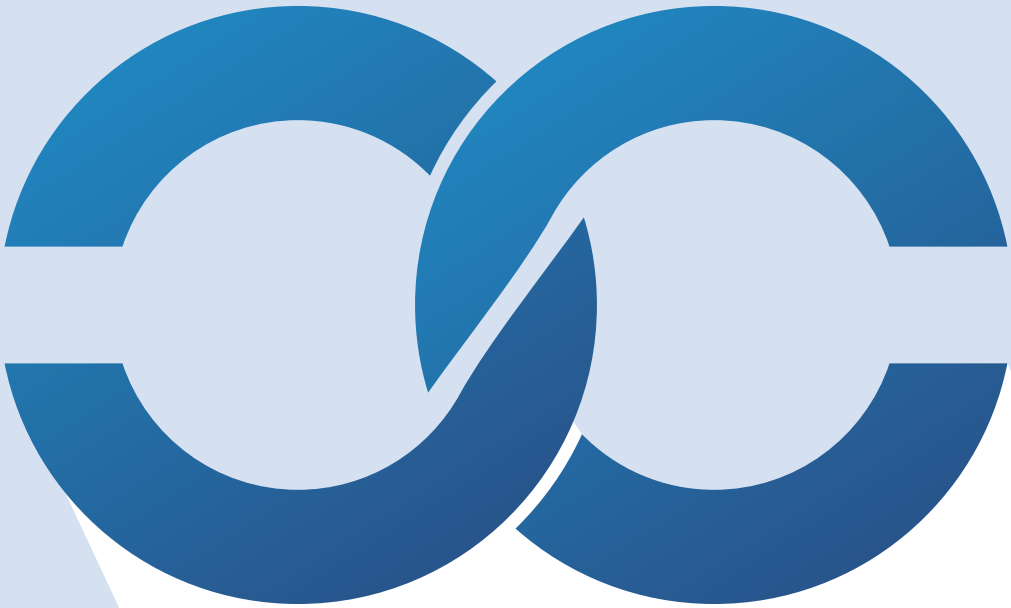
The 'Clinical Connect' newsletter is a commendable initiative by the Fortis group to create a platform for medical practitioners to discuss latest developments in clinical care and medical research.

This bi-monthly newsletter will create a milieu of clinicians and doctors, carving inroads of communication to share knowledge on diverse disciplines. As a neurosurgeon, I am looking forward to interacting with other Medical Council colleagues and doctors of other units of Fortis and 'Clinical Connect' will be an apt medium for it. This free flow of information will help in exchange of ideas and expertise that in turn will strengthen medical science.

The newsletter will bring in views of senior clinicians and will publish credible work of our peers which in turn will inspire all doctors to learn and grow.

I want to thank our editorial team for conceptualizing and publishing this newsletter and hope that the publication, in time, becomes a ready reckoner for all medical queries among doctors and clinicians.





# FROM THE EDITORIAL TEAM

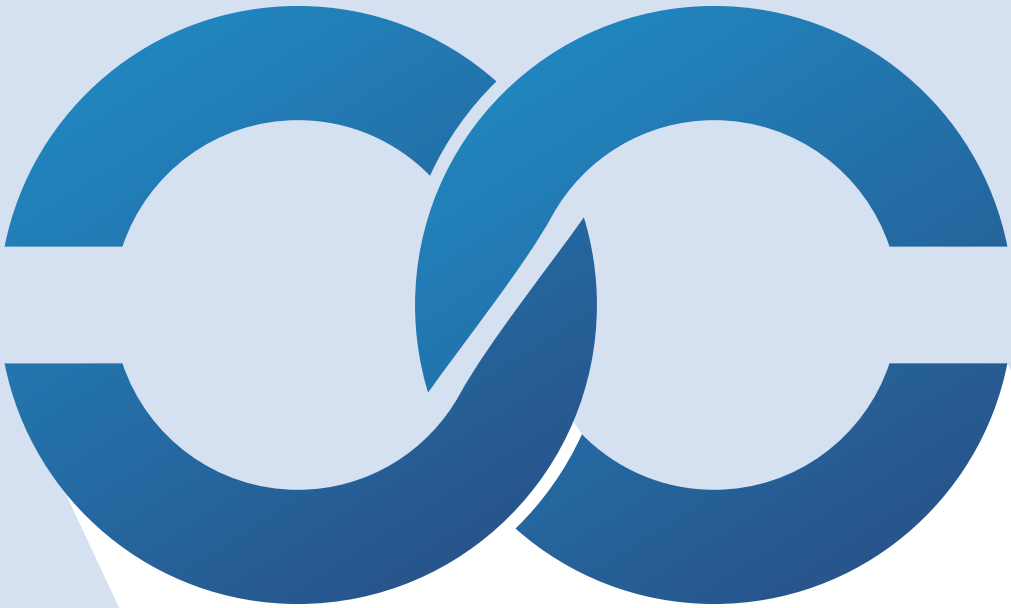
# Message from the Editorial Team

**Dr Sushmita Roychowdhury**
Director-Pulmonology &
Interventional Pulmonology
Fortis Hospital, Anandapur, Kolkata

Our nervous system is the essence of who we are and how we think, behave, perceive, feel, and remember. There has been remarkable progress in understanding this complex branch of science. Diagnosis of a neurological condition is an art, and we look forward to reading a few of the challenging cases and getting updated by the best of neurologists in this country in this issue of Clinical Connect.



Please send your comments, feedback and suggestions to clinical.connect@fortishealthcare.com



# NEURO SCIENCES SPECIALITY COUNCIL

# Neuro Sciences Speciality Council

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Chairperson, HOD and Executive Director - Neurosurery
Fortis Memorial Research Institute, Gurugram

**Dr Rana Patir**
Chairman - Neurosurgery,
Fortis Memorial Research Institute, Gurugram

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Consultant Neurology,
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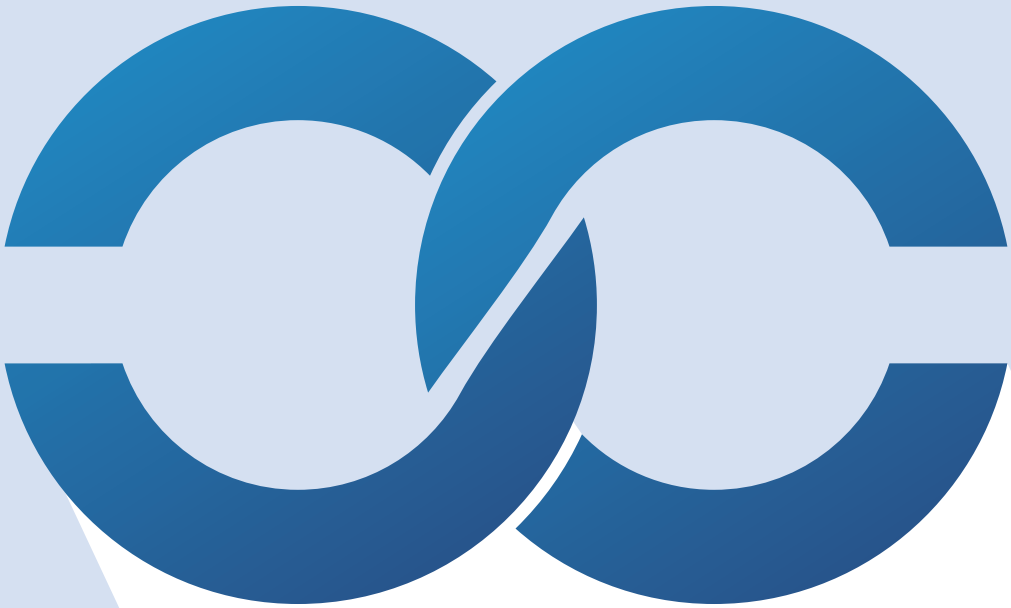
**Dr Rajesh Benny**
Senior Consultant Neurology,
Fortis Hospital, Mulund

**Dr Neetu Ramrakhiani**
Director Neurology,
Fortis Escorts Hospital, Jaipur



# THE WAY WE DO IT AT FORTIS

# World's Largest Spinal Ependymoma - Bone Preserving Innovative Meticulous Surgery to Give a Normal Life to a Young Girl

**Dr Sonal Gupta**
Director & Head - Neurosurgery
Fortis Hospital, Shalimar Bagh,
New Delhi

16-year-old girl presented with a history of back pain for about 18 months and difficulty in walking for a month. Her MRI revealed Intradural tumour extending from D6-S2 (37.35cm)- to the best of my research abilities 28 cm size similar tumour is the largest reported in the world. More than half of her spinal column was full of tumour. (Figure 1, 2)

**Key highlights of surgery:**

1. 12 hours long surgery – Largest Intra-dural ependymoma reported in the world
2. Bone Preserving surgery - We did Open door Laminoplasty technique to preserve normal bone which avoided putting pedicular screws along 14 vertebral levels. Laminoplasty helps in fixing back the normal bone with mini plates and restores near normal anatomy. Most of the spinal tumors are done by removing bone (Laminectomy). More than 3 level laminectomy can produce spinal deformity so those levels are fixed with pedicular screws which leads to rigid spine for rest of the life.
3. Electrophysiological Monitoring - Electrodes are placed to monitor spinal cord and nerve root function. This helps in avoiding any undue manipulation of neural tissue while removing the tumor.
4. Near total Microscopic excision of tumor-except for a small portion which was adherent to spinal cord tissue at D7 level.

Post operatively on 3rd day patient has started walking with support with a Taylor's brace. Her biopsy report is Myxopapillary Ependymoma which is a grade 1 tumour.

Patient will need excellent Neurorehab. and MRI at regular intervals to monitor recurrence. At slightest evidence of tumour growth patient might need radiotherapy too.


Figure 1

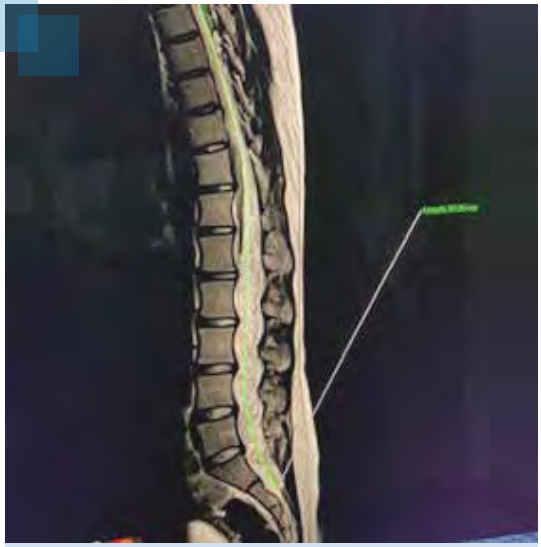


Figure 2

# A Weekend Miracle

**Dr Rajesh Benny**
Senior Consultant - Neurology
Fortis Hospital, Mulund

A 38-year-old male presented to the emergency room (ER) with severe headaches on a Saturday morning of 4 days duration. Over the previous 2 days, he had become progressively drowsy, incoherent, and weak on the left side. On examination in the ER, he was drowsy and had dysarthria with left hemiparesis (grade IV/V). His GCS was 11. His pupillary reflexes and extraocular muscle movements were normal. He had bilateral extensor plantar without any meningeal signs.

He underwent an urgent MRI brain (Figures 1, 2, 3).

Though he had extensive cerebral venous sinus thrombosis (CVT), there was no parenchymal abnormality. He was started on therapeutic anticoagulation with low molecular weight heparin. Over the next 10 hours, his drowsiness progressed (GCS 8). On examination, he had continuous nystagmoid jerks with gaze preference to the left. The left hemiparesis worsened to grade 3 power. He was suspected to have a seizure due to a lesion involving the right frontal eye field. He underwent a repeat MRI which confirmed the right frontal venous edema. (Figure 4)

As he was clinically worsening, it was decided to do a four vessel DSA and attempt intrasinus thrombolysis. A 6F sheath was inserted in the right femoral artery, and another 6F SS was inserted in the right femoral vein. DSA was done using 4F H1 diagnostic catheter over a Terumo wire. There was complete occlusion of the torcula, straight sinus, SSS, inferior sagittal sinus, and bilateral transverse sinus. Cerebral outflow was via cortical veins into the right internal jugular vein. There was prolonged stasis of circulation due to outflow obstruction. (Figure 5)

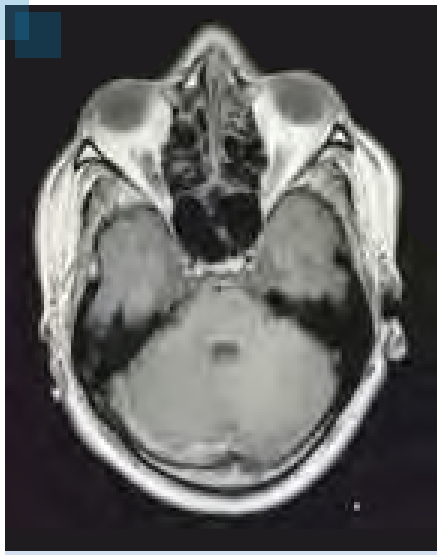
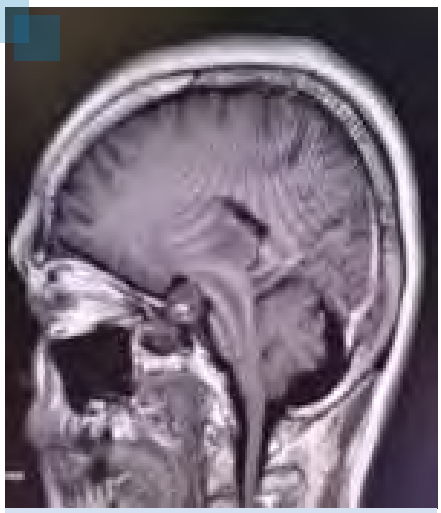


Figure 1



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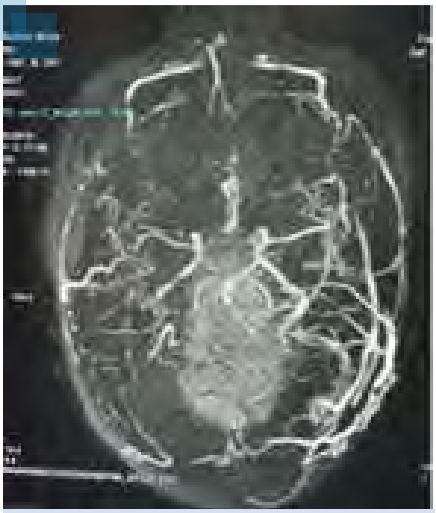


Figure 3

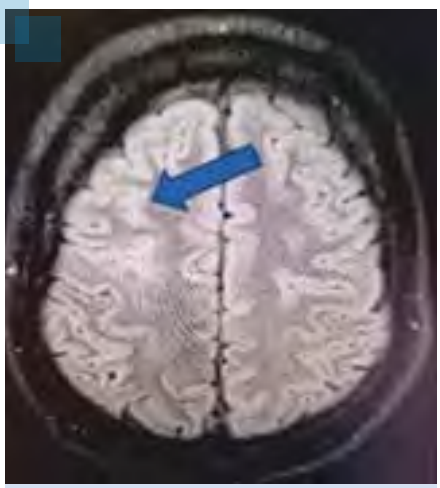


Figure 4

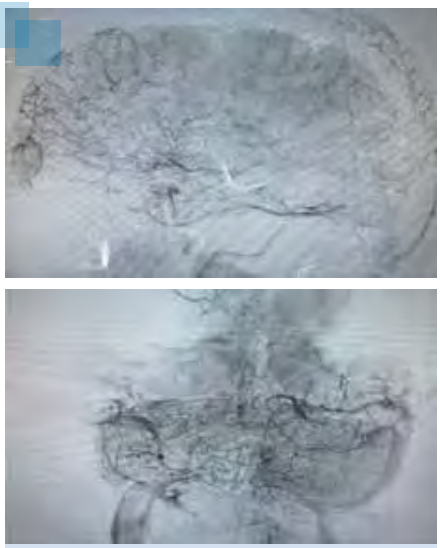


Figure 5

Intrasinus thrombolysis was attempted using Alteplase. A 6F Neuron 70 guiding catheter was navigated via the venous route over the Terumo wire and parked in the right sigmoid sinus. Phenome 27 microcatheter over Traxcess microwire was navigated under roadmap guidance and taken into the torcula. The microcatheter was navigated and taken into the straight sinus. Alteplase (rtPA) 2mg was injected locally. The microcatheter was then navigated under roadmap guidance and taken into the superior sagittal sinus (SSS) (anterior part). The guiding catheter was advanced in the posterior part of the SSS distal to the torcula. (Figure 6)

Figure 6

A large clot was also aspirated.

Alteplase 8mg was injected locally while gradually withdrawing the catheter from the anterior part of SSS to its mid-portion. With the guiding catheter at the posterior part of SSS, infusion of Alteplase at 1.5 ml (1.5mg) per hour was started. The femoral artery and vein sheaths were secured with sterile dressing and infusions in the guiding catheter and microcatheter (Alteplase infusion) were confirmed. The microcatheter was pulled 2 cm every hour for the next 10hours.

The patient was kept sedated and paralyzed to minimize limb movements, agitation, and cerebral edema.

A follow-up DSA was done after 10 hours. There was good recanalization of the straight sinus, SSS, inferior sagittal sinus, torcula, and both the transverse sinuses. There was a significant improvement in the venous outflow (venous drainage was now via both the transverse sinus) and a reduction in the venous stasis. (Figure 7)

The patient was paralyzed medically and ventilated overnight. By the next day, he was conscious without any focal limb deficit and had normal extraocular muscle movement. He was extubated and transferred to the wards the following day. He was discharged on day 5 of admission without any headaches or focal limb deficit.

His pro coagulant workup suggested that he was lupus anticoagulant positive and was discharged on oral anticoagulant (warfarin).

### Intrasinus Thrombolysis In CVT

CVT is an uncommon cause of stroke. In those with encephalopathy, mortality due to CVT can be up to 53%. The Standard of care in CVT is the use of therapeutic anticoagulation which however takes time to recanalize the thrombosed veins (68% of patients achieve partial recanalization in 6 to 10 days). There are no standard guidelines about intrasinus thrombolysis (when to intervene and what thrombolytic to use). Most authors have performed this procedure in those with progressive clinical worsening in spite of therapeutic anticoagulation and in those with deep venous sinus thrombosis.

We were able to alter the natural course of our patient's severe venous sinus thrombosis by identifying subtle neurological signs suggestive of thrombosis progression and urgently.

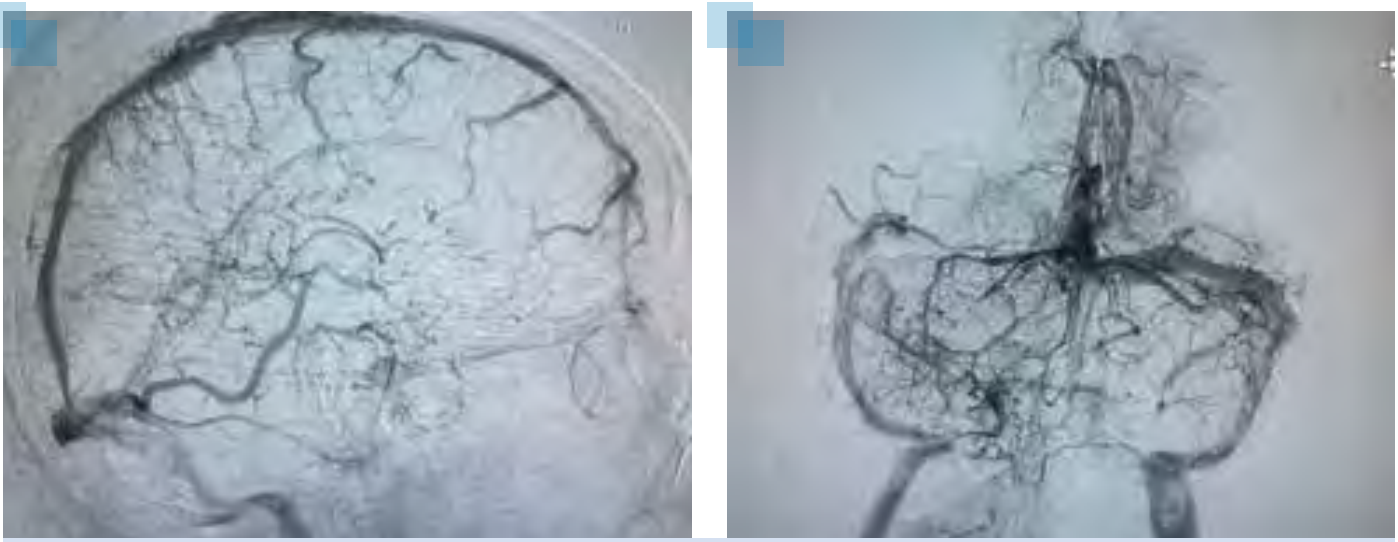


Figure 7

# Prospective Study of Pattern and Outcome of Patients with Ischemic Stroke in Diabetic, Non-diabetic, and Pre-diabetic Individuals in a Hospital Based Setting

**Dr Neetu Ramrakhiani**
Director - Neurology
Fortis Escorts Hospital, Jaipur

## Introduction

Diabetes Mellitus (DM) is identified as a major risk factor in patients with ischemic stroke. It is uncertain whether patients with pre-diabetes carry the same risk and the same has not been systematically explored. Compared with stroke patients without DM, patients with DM had a significantly higher incidence of death or dependency and recurrent stroke at 3 and 6 months after stroke onset.

## Material and Method

This study was conducted on 365 patients to compare the stroke pattern and outcome in pre-diabetic, diabetic, and non-diabetic patients. Successive patients getting admitted to Fortis Escorts Hospital Jaipur with diagnosed acute ischemic stroke were enrolled in this study as per inclusion and exclusion criteria. The data was collected and follow-up was done for 3 months either telephonically or during physical hospital visits.

## Inclusion Criteria

Patients with age >18 years with acute stroke (ischemic) within 2 weeks of stroke onset admitted/ or attending outdoor at FEHJ were included with prior written informed consent. Patients with age <18 years having an intracranial bleed, venous sinus thrombus, and ischemic stroke after two weeks on onset were excluded. Based on the values of HbA1C as per ADA 2019 guidelines patients were categorized as non-diabetic, pre-diabetic, and diabetic.

## Observation and Results

Out of 365 patients, 135patients (36.98%) were non-diabetic, 95 (26.02%) patients were pre-diabetic, and 135 (36.98%) patients were diabetic. In overall study population male distribution was 72.05% and female distribution was 27.95% with p =0.266NS.

The CT scan finding shows acute stroke were 79(79.0%), 50(92%), and 62(73.8%) in non-diabetic, pre-diabetic, and diabetic groups respectively; p=0.231.

In diffusion weighted MRI imaging, 112 (98.2%) non-diabetic patients, 78 (98.7%) pre-diabetic patients, and 102 (95.3%) diabetic patients were diagnosed with acute stroke. (p=0.27).

| Sex | Non-diabetic (N=135) | Pre-diabetic (N=95) | Diabetic (N=135) | Total (N=365) | P Value |
|---|---|---|---|---|---|
| Male | 93 (68.89%) | 66 (69.47%) | 104 (77.04%) | 263 (72.05%) | 0.266NS |
| Female | 42 (31.11%) | 29 (30.53%) | 31 (22.96%) | 102 (27.95%) | |
| Total | 135(100%) | 95(100%) | 135(100%) | 356(100%) | |

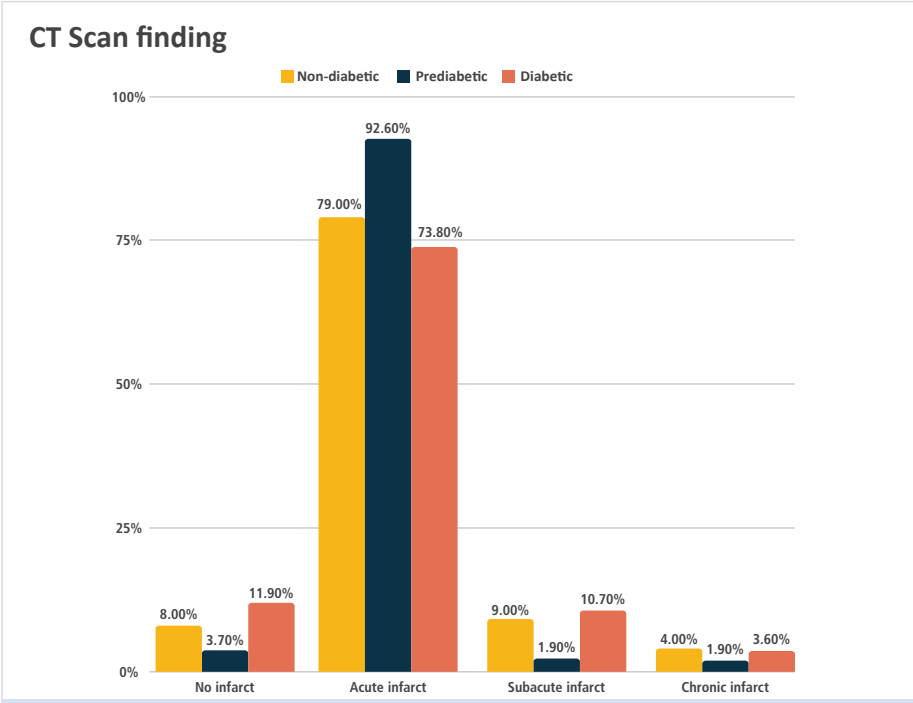



Figure 1

Among non-diabetic, pre-diabetic, and diabetic patients, large artery atherosclerosis had the highest distribution of 37.5% followed by cardioembolism with 21.4% distribution. Diabetics groups are more commonly associated with small vessel occlusion around 23%. P=0.04.

In the mRS scale evaluation the distribution of patients having mRS from 0 to 2 was 28.15% in non-diabetic, 22.22% in pre-diabetic, and 20% in diabetic patients. The mRS from 3 to 5 was reported in 71.85% of non-diabetic, 83.33% of pre-diabetic, and 80% of diabetic patients.

In this study, cardioembolic risk factors are reported in all three groups. Hypertension was observed in 221 patients (60.5%) comprising all three groups followed by diabetes in 127 (60.6%) patients. Prior stroke was found in 54 (14.8%) patients and CAD in 53(14.5%) patients including all three groups. There was a significant association between stroke, hypertension, and diabetes p=<0.001, p=<0.001 followed by CAD p=0.035.

Including non-diabetic, pre-diabetic, and diabetic patients a total of 31 (8.49%) patients were thrombolysed and the distribution was 9 (6.67%) non-diabetic patients, 8 (8.42%) pre-diabetic patients, and 14 (10.37%) diabetic patients. P= 0.479NS.

Other comparable factors among non-diabetic, pre-diabetic, and diabetic were occupation, area of residence, socioeconomic status, family history, lab investigations, dietary pattern, prior medication

NIHSS, and events after discharge up to 3 months of admission. 20 patients (5.4%) were found to be newly diagnosed diabetes as per HbA1C criteria. A longer duration hospital stay was observed in diabetic subjects compared to non-diabetic population. However, no such difference was found in the pre-diabetic group compared to non-

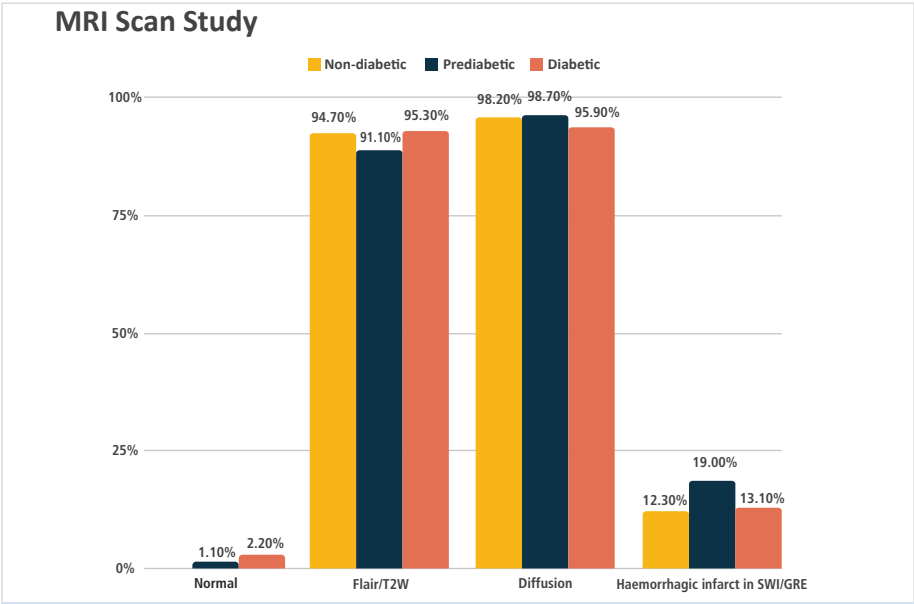
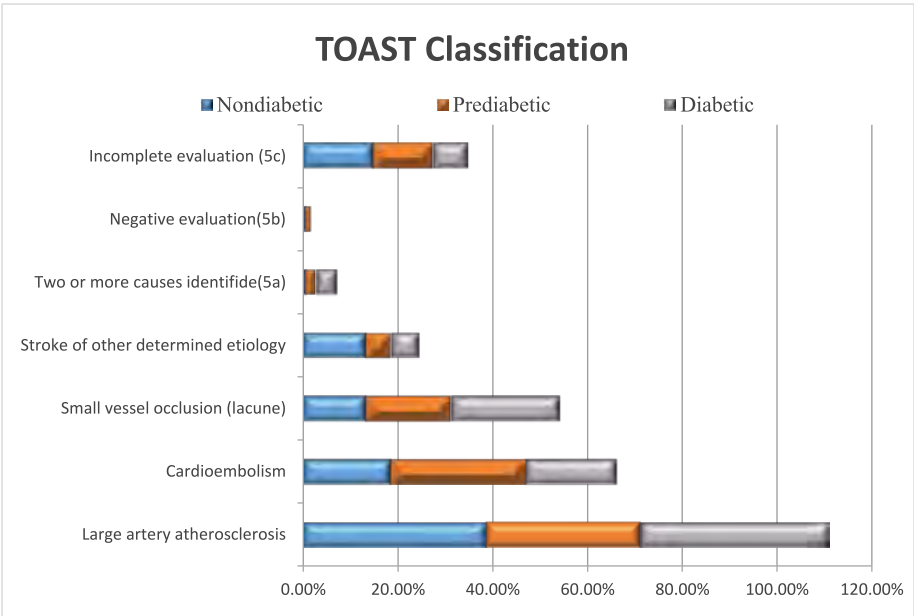



Figure 2

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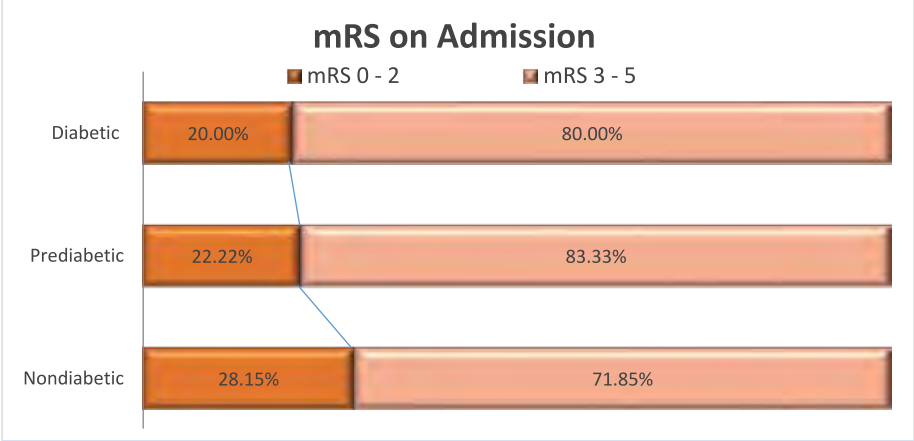


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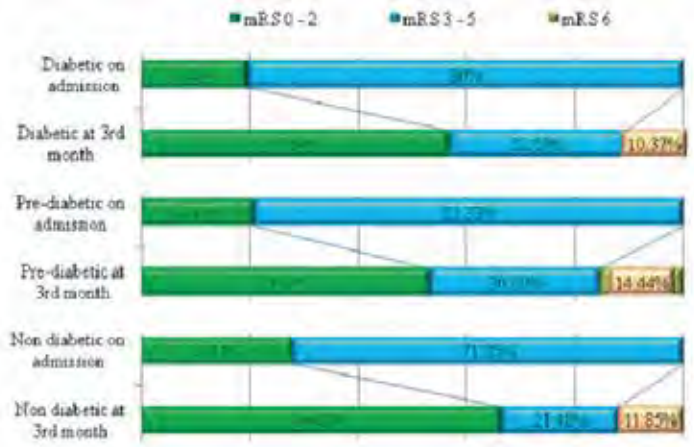


| Events | Nondiabetic (N=135) | Prediabetic (N=95) | Diabetic (N=135) | Total (N=365) | P value |
|---|---|---|---|---|---|
| Death | 16(11.9%) | 13(13.7%) | 14(10.4%) | 43(11.8%) | 0.744 |
| New stroke | 5(4.2%) | 1(1.2%) | 3(2.5%) | 9(2.8%) | 0.43 |
| Seizure | 6(5.0%) | 4(4.8%) | 6(5.0%) | 16(5.0%) | 0.997 |
| Acute coronary syndrome | 2(1.7%) | 3(3.6%) | 2(1.7%) | 7(2.2%) | 0.576 |

Figure 5

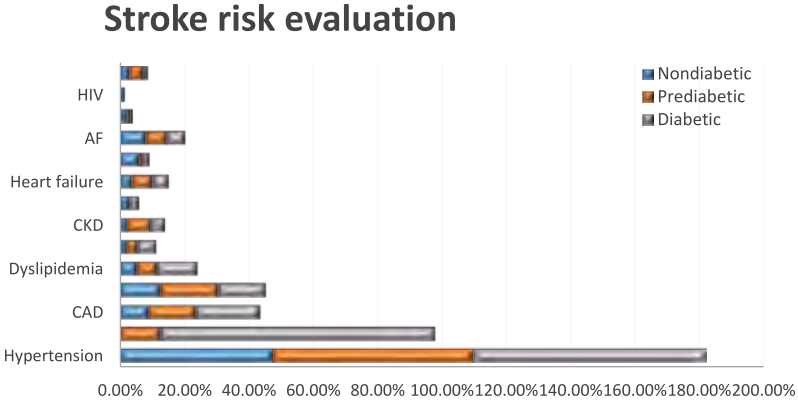


Figure 6

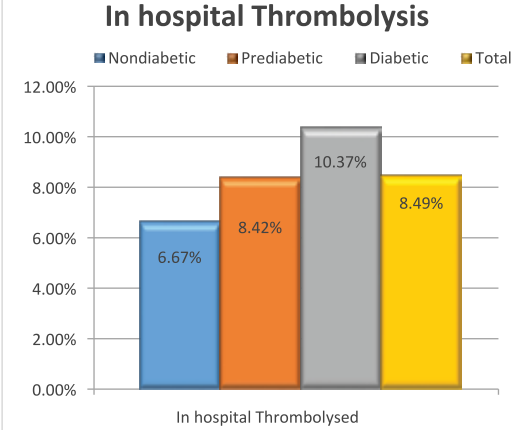


Figure 7

diabetic subgroup. No statistically significant difference was seen in post stroke events.

## Conclusion

After completing the statistical analysis of data, it was observed that the male to female ratio of having a stroke was 72.05:27.95. The mean age of the diabetic group is higher as compared to the non-diabetic and it shows increasing age accelerates the stroke risk. The most common stroke subtype in the diabetic population was large artery atherosclerosis. In the stroke pattern subtype, smaller vessel occlusion was seen in diabetic compared to non-diabetics which was statistically significant. Cardioembolic strokes were found more commonly in the pre-diabetic population compared to the diabetic population. The involvement of MCA in anterior circulation was most commonly found in all three groups. The most common risk factor in diabetic patients compared to non-diabetic and pre-diabetic patients were hypertension and CAD which is statistically significant p=<0.001 & =0.035.

## Recommendation

It is important to screen for glycemia status in patients presenting with acute ischemic stroke. Poor diabetic control leads to an unfavorable outcome in stroke.

This study has been reproduced in part of the thesis by Dr Srinivas Kurupati.

# Improving the Accuracy of DBS and other Stereotactic Neurosurgeries using a 3D-printed Skull for Preoperative Target Practice

**Dr Nishit Sawal**
Consultant - Neurology
Fortis Hospital, Mohali

## Background

Stereotactic Neurosurgery is the technique of accurately reaching a target inside the brain using an external frame of reference. A number of potential sources of error have been identified in frame-based stereotactic neurosurgery; distortion of the frame over time, a difference in the torque used for screw fixation, or simply an inexperienced surgeon. In our centre, we wanted to perform a GPi-DBS using a stereotactic frame that we were previously unfamiliar with. Therefore, we decided to 3D-print the patient's skull, which enclosed within it, GPi targets for pre-operative target practice.

## Methods

CT images were used to make a 3D model of the patient's skull. FGATIR-MRI sequence was used to visualise the GPi. The Bregma was used as a reference point for calculating the coordinates in the MRI images. Using the bregma as a reference, these coordinates were used to create two 5mm targets representing the GPi in the 3D-model of the CT (Figure 1).

## Results

The frame was fixed to the skull, GPi-targets enclosed within the skull were visualised in the pre-operative CT. An RF-electrode was inserted at the intended target and visualised in a postoperative CT (Figure 2). The trajectory was 3.035mm too-medial and 0.486mm too-anterior from the intended target. Fixation screws were adjusted and the accuracy in the subsequent attempt was <0.5mm.

## Conclusions

A 3D-printed skull with surgical targets could help train young functional neurosurgeons. This approach shall also allow neurosurgeons to rapidly transition between the various manufacturers of stereotactic frames, micro-drives and planning stations.

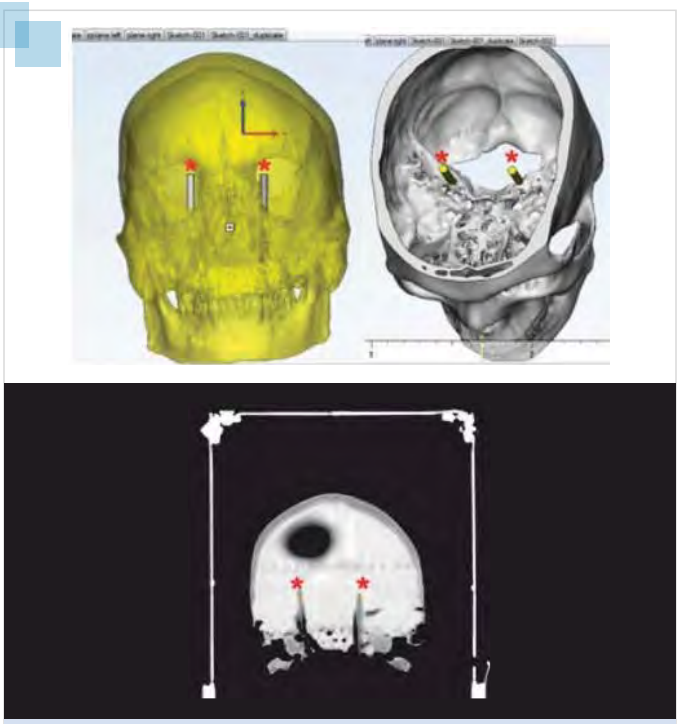

Figure 1: A 3D model of the patient's skull with GPi targets (shown as red asterisks)

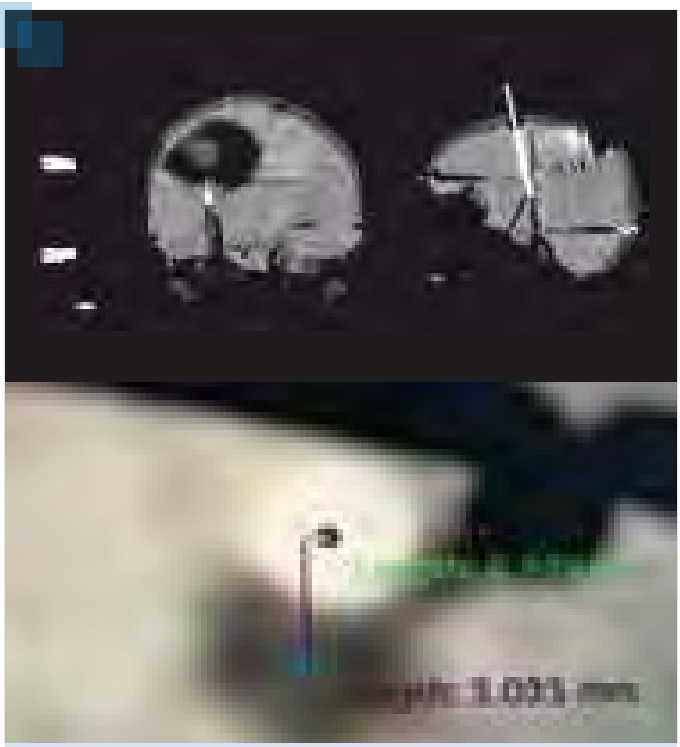

Figure 2

# Minimising Post-Op Micro-Haemorrhages in the Milieu of the DBS Electrode by Using Bipolar Stimulation – An In-vitro Experiment

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## Background

Introducing the Deep Brain Stimulation (DBS) Electrode into the target may cause peri-electrode edema which can present from 6 hours to 120 Days postoperatively, with symptoms ranging from confusional states, neurological deficits to seizures and dyskinetic storms.[1-3] A postulated mechanism of peri - electrode oedema is Microhaemorrhages due to lead implantation.[2] In a prospective study by Borellini et al., all patients who underwent DBS implantation showed asymptomatic peri-lead oedema on postoperative MRI imaging, with 30% having microhaemorrhages.[4] Microhaemorrhages and oedema around the lead can result in postoperative as well as the long term changes in tissue impedance.[5] Therefore, an ideal Lead-Tissue interface, i.e. one with minimal post-operative edema, is essential to prevent post- operative complications as well as to improve long term outcome of stimulation.

Material and Methods

In our in-vitro experiment, two DBS electrodes were introduced into two test tubes, each containing 5 ml of freshly drawn venous blood. One electrode was turned on with a bipolar pattern of stimulation, and the other acted as a control. Clotting time was observed in both the test tubes. The experiment was repeated with increasing increments of stimulation voltage from 0.5 to 4.5 Volts. The experiment was conducted at a constant temperature of 25°C.

## Results

There was a 44.4% decrease in the clotting time with the bipolar stimulation turned on as compared to the control electrode. No change in clotting time was observed after increasing the stimulation voltage beyond 2.0 V. After blood in both test tubes had clotted (at 5 minutes), there was visible clotting on the electrode with the stimulation turned on, while the control electrode was clean.

## Conclusions

Turning on the stimulation significantly reduced the clotting time around the DBS electrode in our in-vitro experiment. We believe these findings will prove to be beneficial in an operative setting. Till date, no evidence-based guidelines exist on when to begin the stimulation after a DBS surgery, with most centres turning on the stimulation after 4 weeks of the implantation. We hypothesise that turning on the DBS electrodes immediately after implantation would help reduce Microhaemorrhages and Oedema by expediting the haemostasis. We believe a bipolar stimulation at 2.0 V for 3 minutes would ensure adequate haemostasis, thereby minimising peri-lead oedema. This would ensure a better lead-tissue interface for future stimulation purposes.

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# A Novel Treatment for Apraxia of Eyelid Opening using a Pulsating Headband: A Single-centre Pilot Study

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## Background

Apraxia of Eyelid Opening (AEO) is characterised by an inability to initiate eyelid opening. It manifests as an isolated condition or along with other neurodegenerative-disorders like Parkinson's Disease (PD) and Progressive Supra-nuclear Palsy. Patients with AEO typically use sensory-tricks (gesteantagoniste) like touching the temporal region or elevating eyelids manually to overcome this impairment. Hirayama et al. (2000) proposed - proprioceptive stimuli to facial-muscles, by wearing skiing/swimming goggles, improved the AEO of patients with PD. Pathogenesis of AEO involves either the Frontalis or the Orbicularis-Oculi muscle. EMG studies showed 80ms-long pulsations stimulated Orbicularis-Oculi and 1000ms stimulated the Frontalis. In our study, we wanted to evaluate the effectiveness of a vibrating-headband that would provide proprioceptive-stimuli to facial muscles for treatment of AEO and compare the effectiveness of 80ms vs. 1000ms-long vibrations.

## Methods

A battery-operated headband with two vibrating-motors was placed just above the patient's eyebrows (Figure 1).

A pilot study was performed on 4-patients with AEO, 1-patient having blepharospasm was excluded (n=3). Using the AEO Scoring System (AEOSS) provided by Ferrazzano et al. (2020), severity of AEO was determined:

1. Without headband
2. With headband-OFF
3. Headband-ON pulsating at 80ms
4. Headband-ON pulsating at 1000ms

## Results

The mean AEO-score without the headband was 6.0±1.7, with the headband-ON at 80ms was 4.7±1.2 and with the headband-ON at 1000ms was 3.3±1.5. Compared to without the headband, there was a 21.6% reduction in AEOSS ($p>0.05$) with the headband-ON vibrating at 80ms and 45.0% reduction when vibrating at 1000ms ($p<0.05$) (Table.1).

## Conclusions

There was a statistically-significant reduction in AEO-score with the headband-ON, vibrating with 1000ms-long pulses, compared to baseline.

Figure 1

| Patient Code | Age/Sex | Diagnosis | Apraxia of Eyelid Opening Scoring System (AEOSS) (out of 10) | | | |
|---|---|---|---|---|---|---|
| | | | Without the Headband | With Headband Turned **OFF** | With Headband Turned **ON** Duration of Pulsations | |
| | | | | | 80ms | 1000ms |
| BR-18 | 66/F | PD* | 8 | 7 | 6 | 5 |
| SB-19 | 71/F | PSP† | 5 | 5 | 4 | 2 |
| BA-19 | 74/F | PSP† | 5 | 4 | 4 | 3 |

**PD: Parkinson's Disease (with STN DBS), †PSP: Progressive Supranuclear Palsy*

Table 1: Apraxia of Eyelid Opening Score of Patient without the headband compared to with the Headband

# Heads - It's Parkinson's; Tails - it's Cancer: Kindred of Parkinsonism and Malignancies

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## Background

Patients with Parkinson's Disease (PD) have increased risk of Melanoma and Prostate Cancer. PARK2, DJ-1 and LRRK2 are genes implicated in both Familial PD and cancer. A 54-year-old male presented to us with generalised stiffness of body for 15 years, starting first from the right lower-limb. His sister also presented with stiffness in her left lower limb for two years, involving right-limb for last 6 months, starting distally and gradually involving whole of limb. We had examined 11 members of a three-generation family, where three individuals had features suggestive of Parkinson's disease and three subjects had a history of cancers. His father had Liver cancer, paternal-aunt had Stomach malignancy and younger sister had a Brain Tumour. The individuals who had a malignancy were spared from PD. (Figure 1)

## Methods

Only two subjects of the eleven examined were wiling for a Genetic analysis, which was done using Whole-Exome-Sequencing (WES).

## Results

In our kindred there is family history of Parkinsonism, there is family history of cancers. Interestingly family members having Parkinsonism are not having malignancies and members having malignancy are not having Parkinsonism. Analysis of WES data allowed us to identify a novel homozygous deleterious non-synonymous variation (D479H) in ZNF837 (Zinc Finger Protein 837) in the index case and his sister.

## Conclusions

This study is first to report any disease association of ZNF837 gene, that too with a neuro degenerative phenotype.

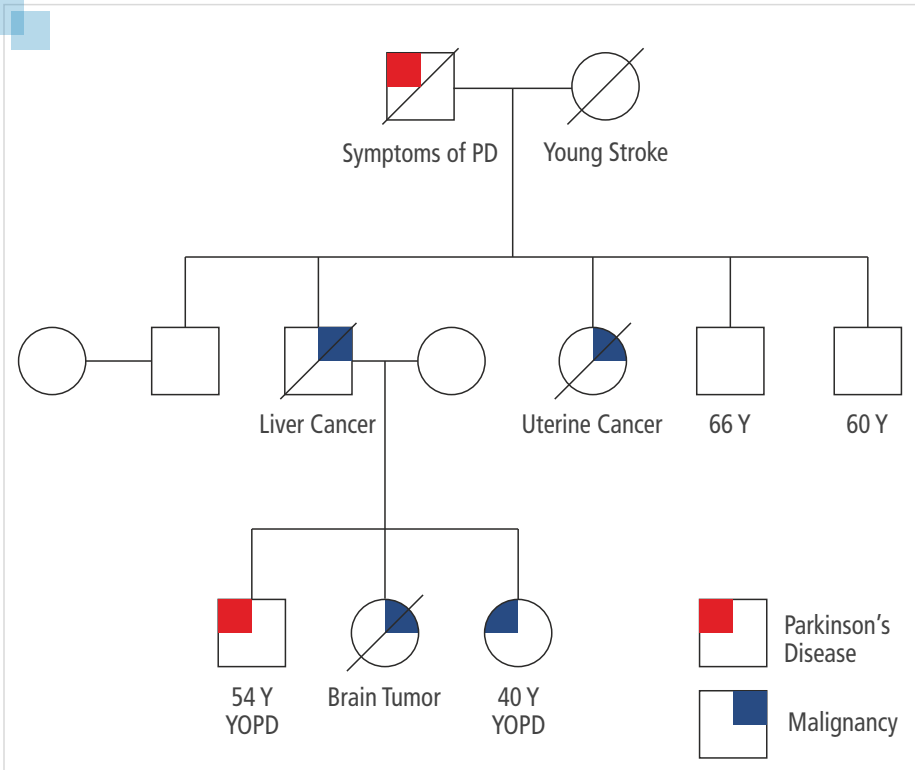


**Figure 1**

# Spine Surgery in a Geriatric Population: Is it really different?

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**Sachin R. Goudihalli, Mandar Patil, Vineet Tanwar**

### Objectives

Spinal degenerative disorders are a major cause of morbidity in the elderly resulting in high dependency. Most of them have a trend to be managed conservatively considering age, comorbidities, and apprehensions of surgical complications. Surgical intervention at early stage with appropriate indications can have better outcomes rather than conservative management in fit patients. The objective of the study is to evaluate the functional outcome in geriatric patients > 60 years who have undergone various spinal procedures for degenerative spine.

### Methods

The study is retrospective, which includes all cases of spinal degenerative disease operated between 2014 and 2016. They were divided into geriatric (>60 years) and non-geriatric cohorts. These include all patients undergoing spinal decompression and/or instrumentation for degenerative disorders of the spine. Patients were interviewed for their functional outcomes in the follow-up period.

### Results

A total of 184 spine cases were operated upon by a single surgeon, out of which a total of 139 cases were operated for the spinal degenerative condition. Forty-eight patients underwent lumbar spinal fusion procedures, 67 underwent non-instrumented lumbar decompression, and 24 patients underwent cervical procedures. These were further divided into 65 geriatric cases and 74 non-geriatric cases.The outcome was assessed with improvement and functional outcomes for spinal disability. Statistical analysis was performed using SPSS 20.

### Conclusion

It is concluded that surgical intervention for spinal problems in geriatric patients is not different from the general population. The outcome is also satisfactory provided, the choice of surgical procedure as per its indication is appropriate. The usual preoperative evaluation for the geriatric age group is very important. The performance status before surgery and the comorbidities have a direct bearing on the outcome in these patients.

### Key Words

Degenerative spine, geriatric, Oswestry, quality of life.

# Neuromonitoring During Neurosurgery – An Emerging Role for Neuroanesthetist

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Intra-Operative Neuro-Monitoring (IONM) is a relatively new technique which has become a standard of care in neurosurgery, wherever functional integrity of neural structures needs to be maintained. Its importance lies in diagnosing injury intraop and in creating a window of opportunity to salvage neural tissues at risk before damage becomes irreversible. With rapid advances in technology and learnings from experience, IONM evolved from the realm of a pure-sciences research modality of the neurophysiologist to a stage where Anaesthesiologists today play an important role. This may be in terms of knowing the patient characteristics, type of surgery and various physiological and pharmacological parameters which will modify the neurophysiological responses, the type of anaesthesia to be used and its impact. To maximise its value, it is essential for the operative team to have a basic understanding of the principles of neuromonitoring and for the anaesthetist to understand how it can be affected by anaesthesia.

Neuromonitoring is used frequently in many procedures today replacing the intraoperative Stagnara wake up test which was described in 1973 by Pierre Stagnara along with anaesthesiologist Mme Vauzelle. The importance of neuromonitoring is such that the American Society of Neurophysiological Monitoring came out with a position statement that evoked potentials are an established practice option for cortical and subcortical mapping and for monitoring during surgeries that may cause injury to the brain, brainstem, spinal cord, or facial nerve.

Different intraoperative neuromonitoring techniques assess the function of the brain, brain stem, spinal cord, cranial nerves, peripheral nerves and neuromuscular junction during the procedure. Neurophysiological monitoring methods include electroencephalography (EEG), electromyography (EMG) and evoked potentials including somatosensory evoked potential (SSEP), motor evoked potential (MEP), visual evoked potential (VEP) and brain stem auditory evoked potential (BAEP).

- Raw EMG (electromyography) which is the most basic parameter, gives the muscles electric activity without any stimulation. It also ensures correct electrode placement into the muscle and of grounding artefacts.
- Triggered EMG: records the response to direct electrical stimulation of nerves or other tissues. A hand-held probe either monopolar or bipolar is used to deliver electric current to the sight of interest.
- MEP or motor evoked potential are electromyographic responses of the peripheral muscles to electrical stimulation of motor cortex; these are electrical signals recorded from neural tissue or muscles following activation of central motor pathways. It identifies the integrity of the cortico-spinal tract.
- SSEP or somatosensory evoked potentials are signals generated by the nervous system in response to sensory stimuli. It assesses the integrity of the ascending sensory tract.
- Brainstem Auditory evoked potential is used to monitor the vestibulocochlear nerve (cranial nerve VIII) and brain stem function.

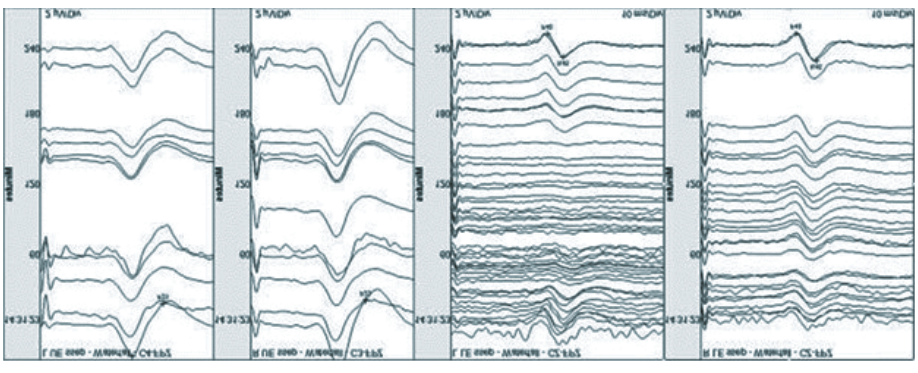

**Figure 1: Figure showing a typical neuromonitoring report**

Monitoring consists of near continuous recording, assessing the ongoing functional integrity of neural pathways. Almost all monitoring is multimodal which means that more than one modality is used during surgery.

Loss of IONM signals or variation from baseline indicates neuronal injury. Multiple factors including anaesthetic agents, blood pressure, body temperature, oxygenation, hypocapnia, anaemia or any positional changes as extreme head position, peripheral nerve compression, spine flexion or extension, and technical faults (as lead failure and electromagnetic interference) should be considered in interpreting the EPs. IV anaesthetics are compatible while inhaled anaesthetics lead to dose dependent suppression of amplitude and increase in latency; muscle relaxants are only used during intubation as they block neurotransmission. Mechanical compression or decrease in blood supply to neural structures also leads to decrease in amplitude and increased latency of signals and are a warning sign of impending neural damage.

The anaesthesiologist plays an important role in evaluating the impact of anaesthesia and physiology on IOM signals and optimising these factors intraoperatively. Good communication with the operative team is imperative and a specifically designed checklist could be helpful in achieving a promising result.

Currently neuromonitoring is conducted by technicians, who are non-medical. It may be worthwhile looking at the prospect of anaesthesiologists privileged to conduct these tests. The reluctance among anaesthesiologists to conduct IONM emanates from a possible medicolegal liability in the event of adverse outcome. In order to put this issue in the right perspective, the neuro-anaesthesia societies must take upon themselves to train its members. In view of the increasing number of IONM, that seems to be the only vista. Academicians and medical administrators ought to come together to put a closure to this sticky subject.

# Science meets Arts and Craft: Fortis Hospital Shalimar Bagh uses 3D Printing Technology to Treat 18-month-old Infant Suffering From 'Trigonocephaly'

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A multi-disciplinary team of doctors at Fortis Hospital Shalimar Bagh treated an 18-month of infant suffering from Trigonocephaly. A cranio-facial surgery was performed to correct the deformity by a team of doctors.

## Clinical Details

"The infant presented with a triangular shaped appearance on the forehead and hypotelorism (a condition in which, the eyes appear closer together than normal). Normally, the bones forming the skull are separated by joints called sutures, which also serve as centres of growth. These sutures fuse at set times after birth, one of the earliest being the suture between frontal bones, the metopic suture which obliterates between 3-9 months of age. An early sutural fusion result in lack of skull growth in a direction perpendicular to the suture. However, the brain is still growing at a rapid rate during this period and needs space to do so. Therefore, the condition needed to be corrected, as it could hamper the neuro-psychological development of the child in addition to the obvious aesthetic defect. (Figure 1)

A cranio- facial approach was needed to correct the deformity. We fed the digital data from the patient's 3DCT scan into the 3D printer to print two rapid protype skull and upper face models, which were exact replicas of patient's skull. We worked on these to determine the exact site, dimensions, and angles of the osteotomies (bony cuts) on the patient's skull. There was no scope for even the smallest of deviations as it could cause the child to go blind, bleed profusely or even suffer from brain damage. To ensure that the actual surgery went off smoothly, we

**Figure 1: Surgery on model created using patient's CT Data**

conducted the mock surgery on the the 3D model of the skull a day prior." During the actual surgery, the next day, we transferred the measurements from the 3D models to patient's skull. We did an entire cranio-facial remodelling by removing the frontal bones (these form the forehead and front part of skull), as well as the orbital bandeau (which forms the lower part of forehead and the upper half of eyeball sockets) while simultaneously ensuring that the brain and eyeballs were protected. We recontoured these bones to the angles and dimensions as per the model surgery and replaced these in corrected position. We used absorbable sutures, miniplates and screws to avoid migration of implants to intracranial or inaccessible locations with later growth of this child. Our work with the two 3D models proved to be fruitful, enabling us to be meticulous in our execution, reducing the operative time and allowing the actual surgery to be precise."

It was a complex surgery which took seven hours. To put an infant under anaesthesia for that long is a challenge. It was an intensive and precise process which would not have been possible without the support of the anaesthesiology team led by Dr Umesh Deshmukh (Director and Head - Anaesthesiology). There was a risk of significant blood loss during the surgery, so multiple blood products were arranged. After the surgery, we shifted the patient to the ICU, where he stayed for 3 days under the care of Dr Amit Singh (Consultant Paediatric Critical Care). Post this period, he was shifted to the ward for 2 days and on the 6 th day he was discharged. Even though I had first met with this patient when he was only two months old, I chose not to the surgery immediately as it would not have yielded the desired results. (Figure 2)

**Figure 2: Pre-op and Post-op Craniosynostosis correction**

# Psychosocial Care for Dementia

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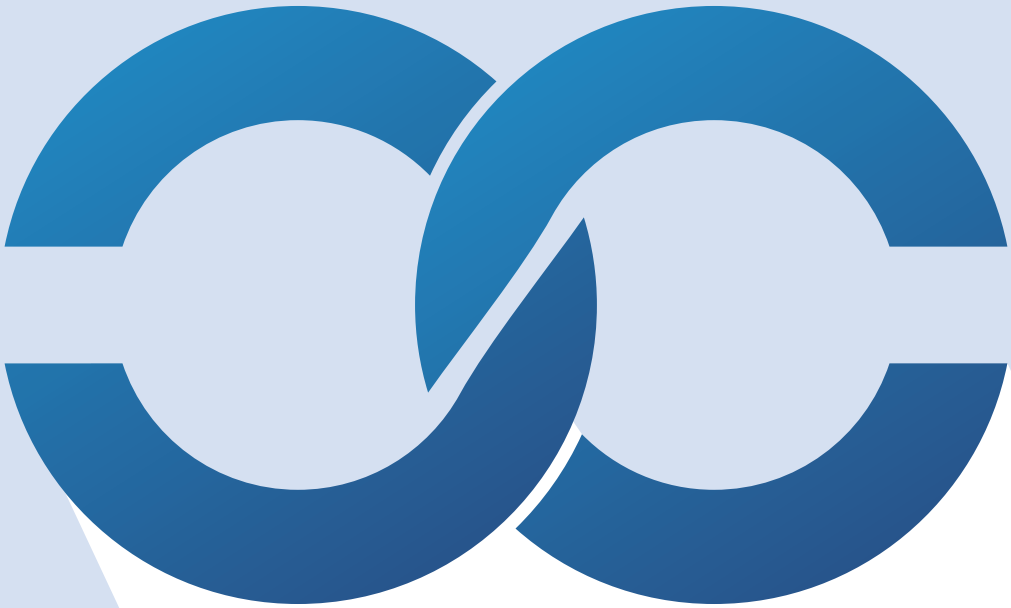
Close to 60 million people in the world are living with dementia, with 10 million new cases added each year. Dementia as an illness affects not just a person's cognitive functioning, but has a deep psychosocial impact on the patient, as well as the family.

In order for patients to cope with their day-to-day life, we need to support them in their activities of daily living like eating, bathing or using the toilet. At this time, be patient and be mindful of their feelings. Respect privacy, encourage independence wherever possible, and make sure to keep talking to them while helping out so they don't feel anxious or caught off guard.

For someone living with dementia, active engagement with their environment is key. Encourage physical activity as much as possible. Outings and walks are helpful. Ensure that they get adequate exposure to sunlight. Play games like sudoku and ludo to both stimulate the mind and strengthen their neurocognitive functioning. Spending time with pets, and looking after plants can also be a calming experience that keeps them engaged. Watching old films and listening to music are known to have emotional benefits and can reduce stress and agitation.

Caring for a loved one with dementia can be an emotionally daunting experience. It's natural to feel stressed, tired or irritable at times. As you care for a loved one with dementia, remember to look after your own self too. Talk to someone and express your feelings. Take time out for yourself whenever you can, and reach out for help when required.

When it comes to looking after someone with dementia, social support is probably the most important factor. Spending time with the person, taking care of them in a respectful, patient and empathetic manner can make all the difference to someone living with this condition.



# CLINICAL CONVERSATIONS

Case Reports

# A Case of Cranial Autonomic Dysfunction Predominant Migraine: Migraine sans Ache

Source:- https://mansapublishers.com/index.php/ijcr/article/view/3007/2468

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Pain is the essential part of migraine headaches along with other features, whereas, cranial autonomic symptoms (CAS)/signs are a predominant and essential part of trigeminal autonomic cephalalgias. What if other features of migraine are present but the pain is absent? One such variation in migraine symptomatology is reported in this case, where the pain was not there but CAS/signs were predominant along with other features of migraine. The possible mechanism for such variation, in this case, maybe desensitization of afferent loop (comprised by nociceptors in extracranial and intracranial pain-sensitive structures supplied by the trigeminal nerve) and direct activation of the trigemino-cervical complex and brainstem structures causing dissociation of pain from the rest of the features of migraine. In this case, as the pain part gets completely dissociated from CAS/signs, hence this condition can be termed as "migraine sans ache." The patient was started on anti-migraine treatment and she responded wonderfully.

Migraine is a common disabling primary headache disorder with recurrent headaches manifesting in attacks lasting from 4 to 72 h. Typical characteristics of the headache are unilateral location, pulsating quality, moderate or severe intensity, aggravation by routine physical activity, and association with nausea and/or photophobia and phonophobia[1]. Migraine attacks can be associated with cranial autonomic symptoms (CAS) like conjunctival injection, lacrimation, nasal congestion, rhinorrhoea, eyelid oedema, forehead and facial sweating, miosis, ptosis, or aural fullness. These CAS are typical and essential features of trigeminal autonomic cephalalgias (TAC) but can be associated with migraine attacks[2,3].

Different variations in migraine symptomology have been described in past and we are here reporting such a variation in this case, where CAS were prominent features rather than pain. Such variation is rare and has not been reported in the literature till now. Detailed symptoms and signs of the patient are discussed and also the possible explanation of this dissociation is discussed by analysing the pathophysiology of migraine especially the role of the trigeminal-cervical complex. Therefore, this case helps us to better understand the intriguing pathophysiology of migraine. Consent was obtained from the patient in question.

## Case Report

A 28-year-old woman presented with episodes of CAS in form of redness and tearing from eyes, bilateral facial and forehead flushing, and bilateral aural fullness without any pain in the head or neck for the last eight months. These episodes were associated with triggers like traveling, hunger, or decreased sleep. Occasional prodromal yawning was present. Photophobia and phonophobia were not present but the nausea was present during few episodes. These CAS evolve gradually, peak over 2–4 h, and subside in 12–24 h. The patient had no pain, but she felt restlessness and discomfort during these episodes. Some activity restriction was also present during these episodes. Though the pain was

not there, she tried analgesics due to the discomfort but were ineffective. She visited our clinic for these CAS and discomfort during episodes. No menstrual associations with CAS were present. Stressors were absent. Psychiatric co-morbidities screened by the Patient Health Questionnaire showed negative results. No history of any eye inflammatory conditions such as uveitis or conjunctivitis in the past. No history of any connective tissue or autoimmune disorder in past. Family history for headaches was negative.The patient was afebrile, her blood pressure was 120/70 mm of Hg, with a normal respiratory, cardiac and abdominal system on general physical examination. Neurological examination including higher mental functions, cranial nerves, motor, sensory, cerebellar, and extrapyramidal system was within normal limits.

Local eye examination during attacks shows conjunctival injection and watering in both eyes. Bilateral forehead flushing was also present during attacks.

On investigation, the patient's blood counts, electrolytes, renal function, and liver function tests were within normal range. Her erythrocyte sedimentation rate was 9 mm/h. Thyroid function test, Vitamin B12, and Vitamin D levels were within normal range. Autoimmune and connective tissue disorder workup (ANA, c-ANCA, p-ANCA, and APLA) was negative. Magnetic resonance (MR) imaging brain with MR angiography of the brain and neck vessels did not show any abnormal parenchymal or vascular lesions.

Although she was not fulfilling the criteria of episodic migraine without aura as she was having characteristic features of migraine other than pain, a diagnosis of "migraine without aura with predominant CAS" was considered. The patient was started on propranolol 20 mg twice daily for prophylactic treatment and naproxen sodium 500 mg with domperidone 10 mg combination for acute termination of CAS. She was advised to follow-up with a headache diary which was pointing out CAS in terms of onset, severity, laterality, and frequency. Her CAS with other features such as prodromal yawning, nausea, and restlessness gradually improved and completely subsided over 3–4 months.

## Discussion

One or more unilateral or bilateral CAS can be present in around 27–73% of patients with migraine [4-7]. However, CAS themselves does not define migraine, and unilateral CAS are characteristics of TAC [1]. In this case, episodic bilateral long-lasting CAS were present with nausea, restlessness, and some activity restriction. It can be inferred that, in this case of migraine, the pain part gets dissociated leaving behind a significant CAS part and few other associated features such as nausea and activity restriction. However, these CAS predominant episodes do not fulfil current the International Classification of Headache Disorders-3 classification criteria for migraine or any other headache of current classification. As these CAS episodes were episodic, bilateral, and of long duration, these cannot be justified in the TAC group also. As the pain was absent with other features of migraine, therefore "migraine sans ache" terminology can be used for this type of headache. Other disorders where CAS is found to be associated with headaches are few secondary headaches like pituitary dysfunction, intracranial aneurysm, and sinus headaches [8]. These secondary headaches were ruled out by brain imaging. Lesions in the higher autonomic centre of the brain and any bilateral lesions in the sympathetic and parasympathetic nervous systems were also ruled out.

In this case, to properly understand the phenomenology, we need to understand the pathophysiology of CAS in different headache disorders. Sensory oversensitivity causes activation of nociceptors in extracranial and intracranial pain-sensitive structures supplied by the trigeminal nerve causing pain [9].

Signals from the trigeminal nerve traverse trigeminal ganglion and reach the trigeminal nucleus caudalis and the upper cervical spinal levels forming a trigemino-cervical complex. From the trigemino-cervical complex through genetically sensitized pathways signals reach to brainstem, medullary, diencephalic, hypothalamic, and thalamic areas, and finally to the parasympathetic superior salivatory nucleus (SSN) in the pons, activation of which causes CAS. Therefore, pain in the trigeminal nerve and upper cervical spinal nerves supplied area of the head and neck is a predominant feature with CAS depending on the involvement of brainstem sensitization and activation. This phenomenology is present in most migraine patients.

Rozen TD described such dissociation of pain and CAS in 2 patients with long-lasting autonomic symptoms with hemicranias (LASH) where CAS exists beyond pain and lasts longer than pain [10,11]. But in LASH, pain and CAS were hemicranial and LASH was described as indomethacin responsive TAC. Toribio-Díaz ME also described 10 cases with prolonged eyelid edema (one of CAS) accompanied with and beyond the pain of migraine [12]. He considered it as an accompaniment of migraine and it was improved with anti-migraine treatment. Our case is unique and different in many aspects. Here, the pain part gets completely dissociated from CAS, hence such a condition can be termed as "migraine sans ache." The presumed cause of

this dissociation of pain from the rest part of migraine may be desensitization of afferent loop and direct involvement of the trigemino-cervical complex or brainstem structures (without the participation of nociceptors) causing reflex activation of SSN in pons producing CAS without pain.

## Conclusion

This case helps us to better understand the pathophysiology of migraine and possible variations in phenomenology in migraine. Therefore, migraine is still a mysterious disease, and variations in phenomenology are possible. We need to identify and report these variations more frequently so that we can better understand this very common disease.

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# A 46-year-old has a New Lease of Life Following a Major Spine Surgery for Severe Cervical Spinal Cord Compression

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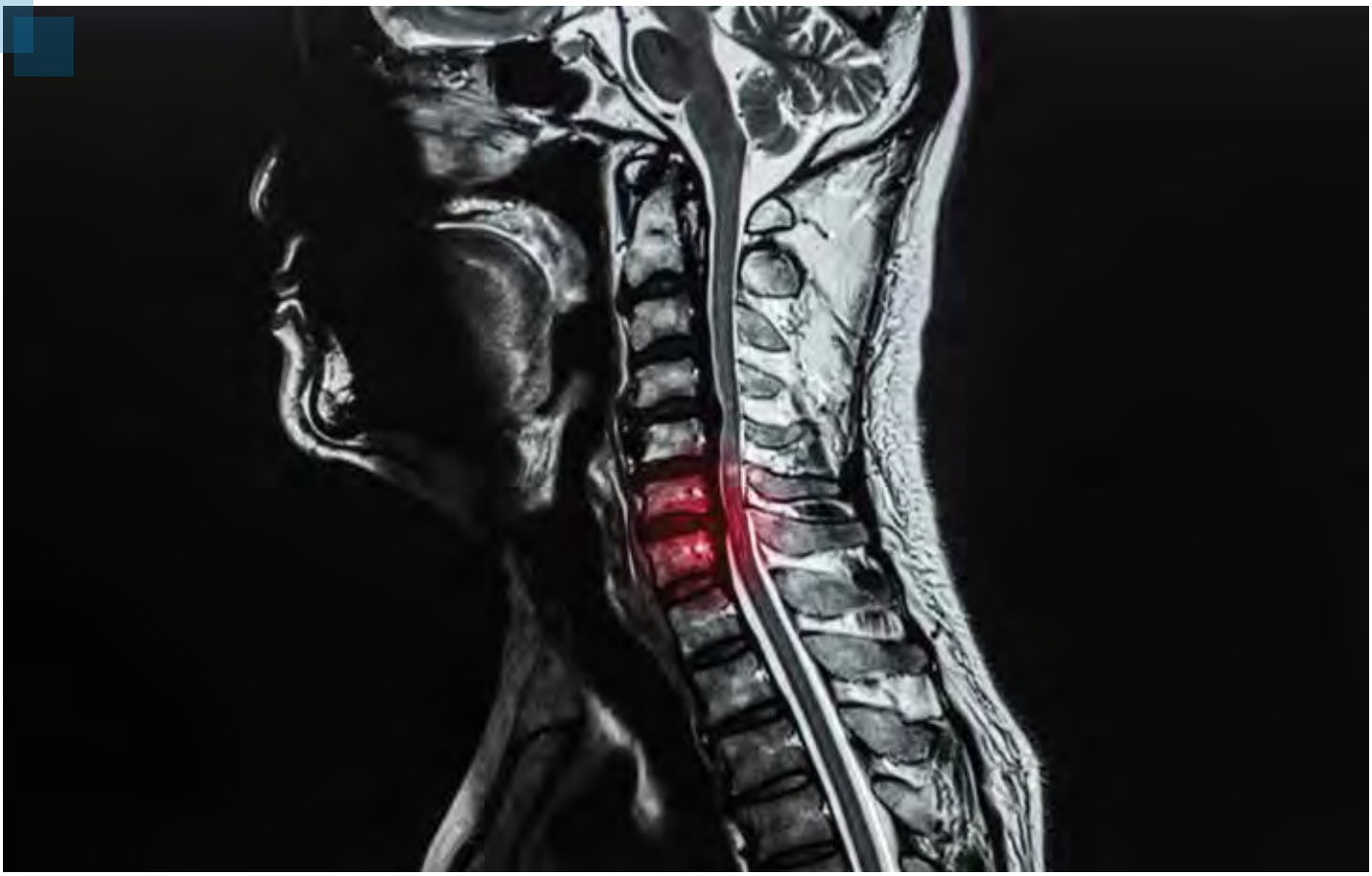
Myelopathy is a serious problem affecting the spinal cord, which if left untreated could lead to permanent neurological dysfunction. A 46-year-old male presented with complaints of gait instability, weakness of hands, difficulty in buttoning his shirt, loss of dexterity, and frequent falls for the past one year. Recently he started developing bladder and bowel disturbances. Being a young chronic alcoholic, most of his clinical symptoms were attributed towards his personal habits by his family physician and as things worsened, before he presented to us.

Diagnosing patients with cervical cord myelopathy in a timely manner can be challenging due to varying clinical presentation and absence of pathognomonic findings at the early stages. The patient gradually lost his ability to perform activities of daily living and was bed ridden and unable to stand when he first met us. On examination he had severe wasting of the intrinsic muscles of both hands and the deep tendon reflexes were exaggerated in both the upper and lower limbs. With bilateral hyperreflexia, Babinski reflex, and positive bilateral Hoffman reflex, a clinical diagnosis of myelopathy was established.

Sagittal section (a) from a T2-weighted Magnetic resonance imaging revealed spinal cord compression at multiple levels with most severe canal compromise at C3-C4. Spinal Cord myelomalacia extends from C3 to C6 and Axial sequence (b) MRI showed severe spinal cord thinning at C3-C4. Upper cervical spinal cord compression is challenging due to involvement of the phrenic nerve which receives its main input from the fourth (C4), and partly the third (C3) and fifth (C5), cervical nerve roots. Additionally, patients with chronic cord compression, long standing disability and higher mJOA



scores do not improve significantly following surgical decompression.

This 46-year-old gentleman and his family members were explained about the need for early surgical intervention mainly to prevent progression of myelopathy and further disability. Considering the multilevel cervical canal stenosis, a posterior spine surgery with cervical spine stabilization from C3 to C6 along with laminectomy was planned. In critical cervical canal stenosis, there are several reports of neurological deterioration during intubation and surgical positioning. The spinal cord has a significant watershed area causing anterior spinal cord ischemia even during slightest alteration in neck position in such cases. It is important to either use awake fiber optic intubation or Multimodal Intraoperative Neuro Monitoring (ION M) for preventing untoward incidents during positioning. We had used both Somatosensory evoked potentials and Motor evoked potentials (MEPs) in this surgery, which required total intravenous anesthesia with propofol infusion for IONM and baseline evaluation was done before and after intubation as well as following prone positioning. While both upper limbs (C5, C6 and C8) had good amplitude all the time, due to severe cervical stenosis no MEPs were recordable even during baseline and the same was conveyed to the relatives before positioning for medicolegal reasons. Though SSEPs remained normal throughout the procedure, it's use in IONM as a single modality is limited. We performed a C3 to C6 posterior cervical stabilization using lateral mass screws (c) followed by a cervical laminectomy (d) to decompress the spinal cord. Additionally, C5 foraminal decompression was performed to prevent C5 palsy. (Figure 1)

Considering the higher level and severity of cervical cord compressive lesion, this patient was gradually weaned off ventilation post operatively and extubated on the next day. He was put on an enhanced recovery path and spinal cord injury rehabilitation program was initiated immediately which included ambulation (e) on the third post-operative day. There has been a good recovery of neurological function and his mJOA score has improved from 9 to 14 over two months with good recovery in bowel and bladder function. Being ambulant on his own this patient is all set to start a new business in the few months. Fortis Vadapalani is now all set to take up challenging spine cases as witnessed by the joint collaborative effort by the team of spine surgeon, intensivist, anaesthesiologist, and physiotherapist.

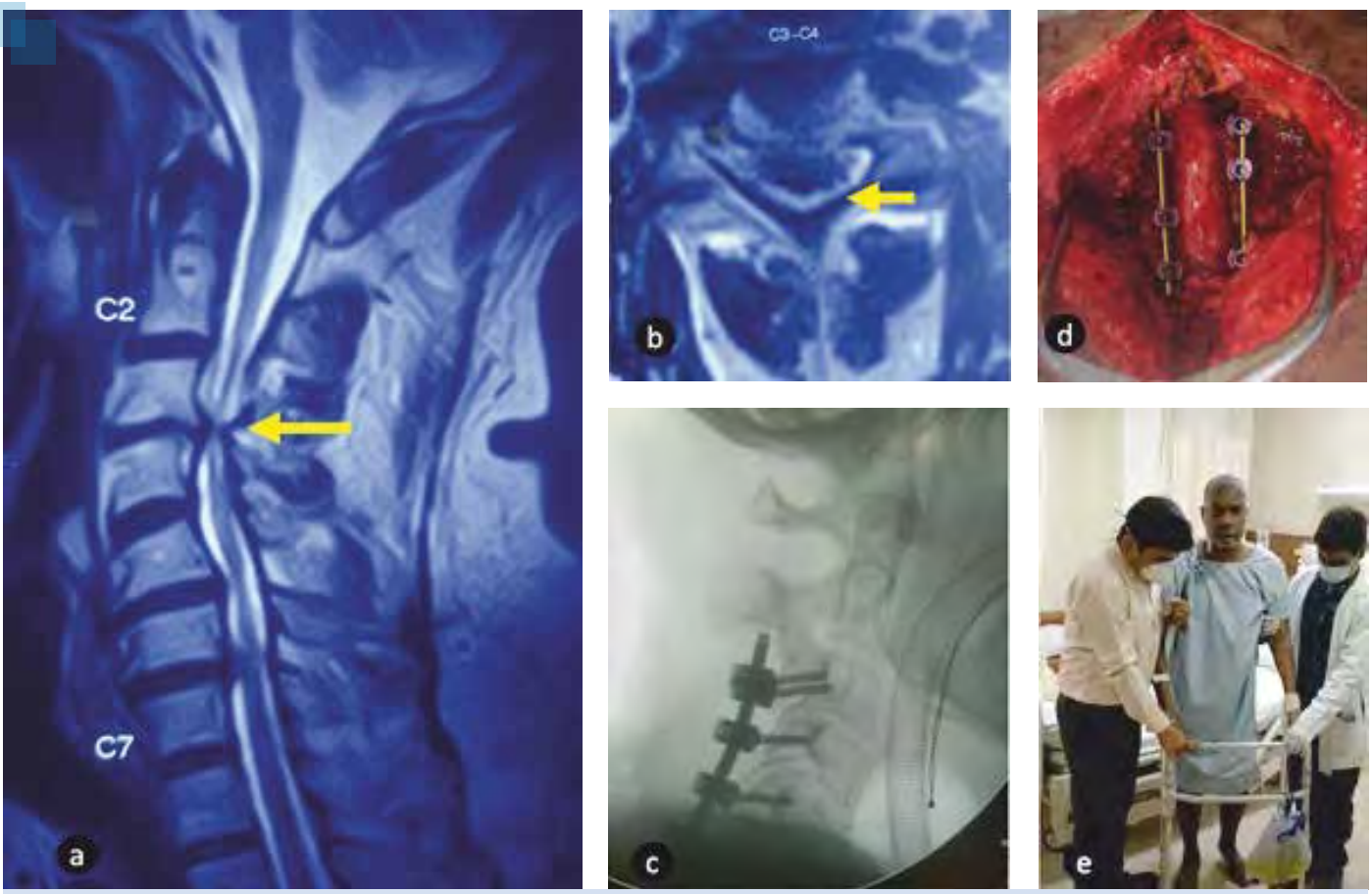



Figure 1

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# Orthostatic Myoclonic Jerks in a Case of Hashimoto's Encephalopathy

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Hashimoto's encephalopathy (HE) is an uncommon syndrome with the characteristic triad of positive antithyroid antibodies (most commonly antibodies to thyroid peroxidase), response to steroids, and clinical picture presenting either as stroke-like pattern of focal neurological deficit or slowly progressive cognitive impairment. Myoclonus or tremors, seizures, and psychosis are other associated features which can be seen in HE. Herein, we report a girl with an uncommon presentation of orthostatic axial and myoclonic jerks in bilateral lower limbs in a case of HE.

## Case Presentation

A 16-year-old girl presented to the clinic with complaints of inability to walk for almost 2 years due to jerky movements in both lower limbs while standing and severe imbalance while walking. These abnormal jerky movements were aggravated by putting load over her limbs and completely subsiding while sleeping or at rest. Her symptoms were subacute in onset and progressed over few weeks, hampering her movements completely. Before our assessment, patient was treated as psychosis and conversion disorder, but there was no improvement in her condition.

On examination, she had bilateral lower limb and axial myoclonic jerks which increased on standing, suggesting orthostatic nature of these jerks. These myoclonic jerks led to severe postural imbalance so much so, that she had to take support of others to walk. No motor weakness or sensory deficits were present. Reflexes were normal. Mild lower limb cogwheel rigidity was present on examination. Rest of physical examination was unremarkable. Her routine blood investigations were normal. Magnetic resonance imaging of the brain and spinal cord was normal. Electroencephalography and nerve conduction studies were normal. Cerebrospinal fluid (CSF) examination showed mild increase in proteins, and rest was normal. Antinuclear anti- bodies were negative. Serum copper and ceruloplasmin levels were normal. Autoimmune encephalitis panel in CSF and serum were negative. However, she had T3 of 54 ng/dL (80–200) and T4 of 4.1 µg/dL (5.10–14.10) levels with elevated thyroid-stimulating hormone 16.4 µIU/mL (0.27– 4.20). Antithyroid peroxidase (anti-TPO) antibodies were strongly positive (1,200 IU/mL [0–8]). With this clinical profile, positive anti-TPO antibodies, normal metabolic, infectious, structural and other autoimmune parameters, HE was diagnosed and pulse intravenous steroid therapy (1 g intravenous pulse methylprednisolone once daily for 5 days) was started with levothyroxine supplementation, followed later by oral steroids (1 mg/kg/d for 2 weeks) in tapering doses.

Following 5 days' course of intravenous steroids, patient started responding slowly after third dose, with gradual and steady improvement. Complete clinical resolution was attained with 3 weeks of therapy, which later persisted on oral steroids. Finally, the patient was able to walk without any support with a total treatment duration of 3 to 4 weeks.

## Discussion

HE is a distinctive neurological disorder with specific clinical features in conjunction with high antithyroid antibodies. However, antithyroid antibodies can be seen in the normal population also. Recent articles by Mattozzi et al2 and Delgado-García and Balint3 discussed about the controversies surrounding HE. With recent advancements and new revelations in immunology and autoimmune neurological disorders, in future, this entity may be amalgamated with other disorders, but for the time being, any clinical syndrome with very high anti-TPO antibodies, characteristic clinical features, and good response to steroids is regarded as HE.

Since the description by Brain et al,4 many new symptoms have been added to the spectrum of HE. Extrapyramidal system involvement leading to varied movement disorders is now being recognized in HE, including myoclonus, tremors, ataxia, dystonia, chorea, tics, palatal tremors, paroxysmal kinesigenic dyskinesia, myorrhythmia, and parkinsonism.5–7 The index patient had myoclonic jerks, but they were pre- dominantly present in lower limbs and were noted only instanding posture, suggestive of orthostatic nature of these myoclonic jerks.

Orthostatic myoclonus term was first introduced by Glass et al8 in 2007 from Mayo Clinic as a cause of gait impairment in neurodegenerative disorders. Since then, several aetiologies have been reported for orthostatic myoclonus, such as Parkinson's disease and atypical parkinsonism, dementia with Lewy bodies, Alzheimer's disease, mild cognitive impairment, normal pressure hydrocephalus, cerebral amyloid angiopathy, mild small vessel cerebral disease, post radio- therapy of resected frontal brain tumour, osmotic demyelination following rapid sodium correction, Lance–Adams' syndrome, and

contact in-associated protein-2 positive au- to immune encephalitis.

The exact pathophysiology of orthostatic myoclonus and why jerks only occur while standing in these varied disorders is still not known, but the most plausible reason is the presence of a subcortical generator. A possibility of ponto-cerebellothalamocortical network involvement is also likely but further investigations such as functional imaging studies in such patients may clearly elucidate the exact pathophysiology of this disorder.

Cortical and subcortical myoclonus have been previously described in HE, but orthostatic myoclonic jerks have not been hitherto reported. Because of these orthostatic myoclonic jerks, the index patient had postural imbalance and she was not able to stand or walk without support. These abnormal movements cannot be categorized as orthostatic tremors as they persisted on walking. The current case illustrates the fact that orthostatic myoclonic jerks can be a presenting rare movement disorder associated with HE and can be treated with standard steroid treatment.

## Conflict of Interest

None declared.

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# Flow-diverter in a Ruptured Supra Clinoid Aneurysm

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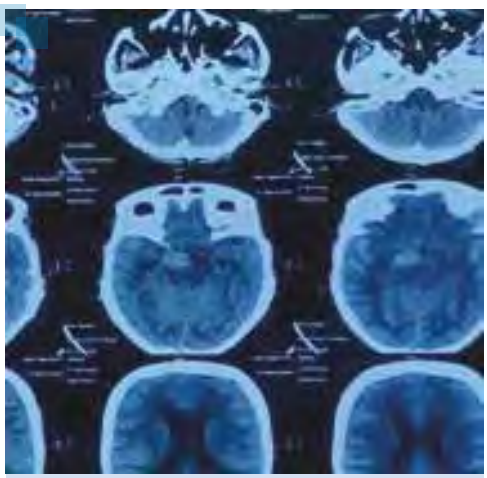

Figure 1: Revised Fisher Grade

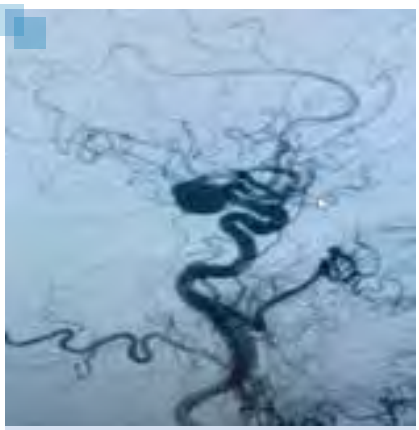

Figure 2:
Large right supra clinoid- anterior choroidal aneurysm; lateral view

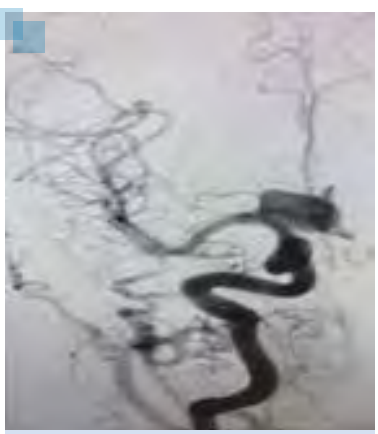

Figure 3: Large right supra clinoid-anterior choroidal aneurysm; AP view

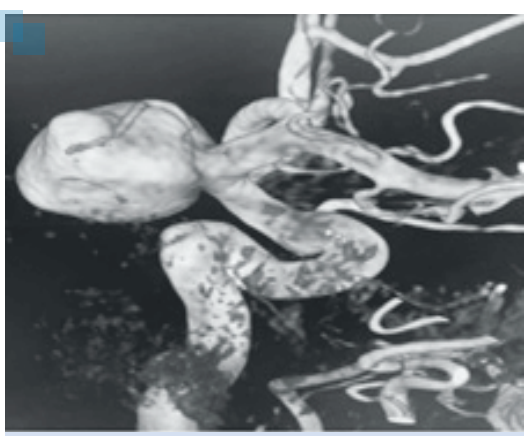

Figure 4: DSA 3D reconstructed image showing the large right supra clinoid-anterior choroida

50 yrs. old lady presented to us with sudden onset headache, vomiting and loss of consciousness. Patient had been admitted to another hospital where she had been preliminary investigated with working diagnosis of SAH with ruptured right supra-clinoid aneurysm.

At the time of admission, the SAH WFNS grading was 2 and revised Fisher grade 3. Patient had been on irregular treatment for hypertension. (Figure 1,2)

A fresh CT image at our hospital revealed SAH with a hyper dense shadow on right side just next to anterior clinoid abutting the peduncle of brain stem suggestive of a large intra cranial aneurysm. The CTA images which were not readily available to us at the time of admission were of poor quality with 3D reconstructed images not showing any aneurysm but native CTA images confirming the diagnosis of a large right supra-clinoid aneurysm.

Relatives were counselled and patient taken for DSA under GA, which confirmed the presence of large right supra-clinoid anterior choroidal aneurysm. (Figure 3,4)

Various options were discussed within the team vis a vis surgical clipping versus endo vascular treatment (EVT).

Relatives were willing for EVT with surgical clipping to be reserved as a secondary rescue procedure.

Patient was loaded with dual antiplatelets on table and a Flow Divertor stent deployed across the neck of the aneurysm, at the same time the aneurysm was loosely packed with coils, thereby completely securing the aneurysm. (Figure 5,6)

Patient did well post operatively and was discharged 7th post procedure day.

Patient was doing exceptionally well in follow up, with no gross neurological deficit.

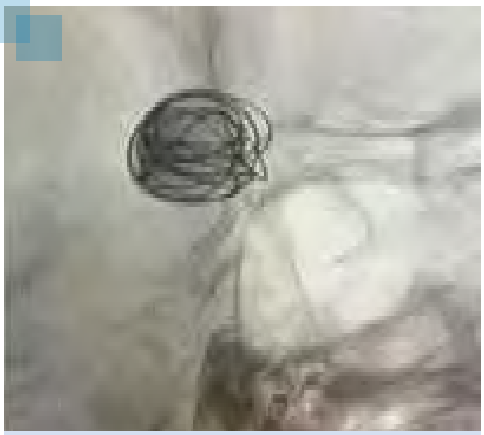

**Figure 5: Flow Divertor deployed across the neck with loosely packed coil mass in the aneurysm**

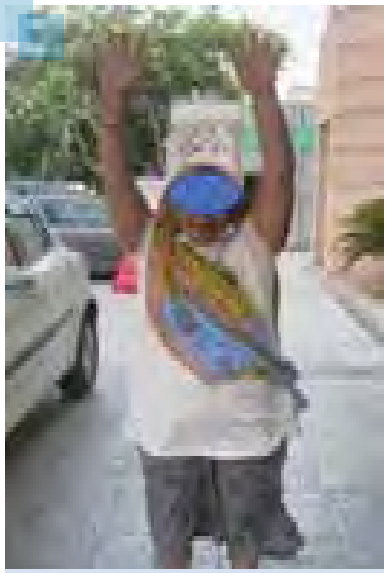

**Figure 6:**
**Patient in first follow up**

# Nocardia Brain Abscess: A Sinister Aetiology

**Source:- https://pmj.bmj.com/content/98/e1/e30**

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A 42-year-old man presented with episodic headaches for the past 2 months, accompanied by palinopsia (visual perseveration). Medical history was significant for sarcoidosis, necessitating prednisolone use for the past 6 months. The patient had normal vitals, fundus and neurologic examination except for a weak right-hand grip. MRI of the brain revealed multiple conglomerated brain abscesses in the left occipito-parietal region (figure 1A). Contemplating a tubercular aetiology, antitubercular therapy (ATT), levetiracetam and dexamethasone were started. However, after 3days, the patient became drowsy and developed right-sided hemiparesis. Re-evaluation showed papilledema and increased size of abscesses (Figure 1B). The largest abscess was immediately surgically drained (Figure 1C, D). Microscopic examination and pus culture showed modified acid-fast beaded hyphae suggesting Nocardia farcinica (Figure 1E, F). ATT was withdrawn and intravenous trimethoprim/ sulfamethoxazole (TMP-SMX) (5mg/kg/day eight hourly) and imipenem (500mg six hourly) were initiated. After 4 weeks, the patient was continued on oral TMP-SMX only. He responded remarkably, with significant radiological and clinical resolution following 2 months' therapy.

Headaches with red flag signs such as acute-onset, seizures, fever, visual disturbances or palinopsia warrant a detailed investigation for underlying aetiology. Opportunistic infections such as Nocardia brain abscess must be suspected in immuno-compromised patients even without fever. Nocardia is a Gram-positive actinomycete that causes suppurative infections including craniocerebral involvement (25%), but brain abscess is rare (2%). Neuroimaging followed by microbiological testing clinches the diagnosis in most cases. Management entails combined surgical and medical treatment with 3–6 weeks intravenous followed by prolonged oral antibiotic therapy for 6–12months. (Figure 1)

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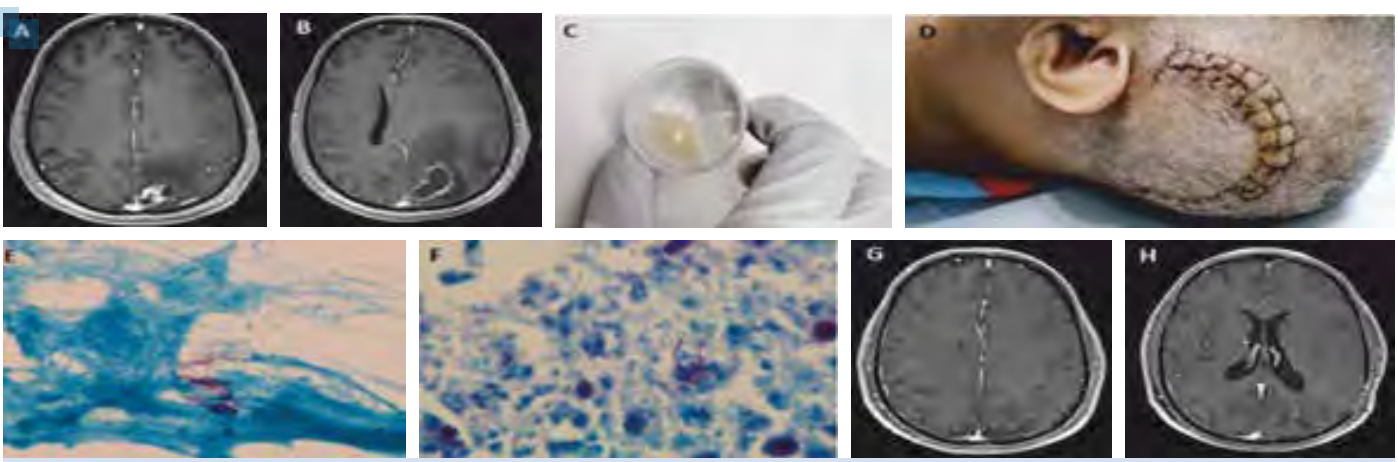

**Figure 1: Panel of images depicting (A) contrast enhanced MRI of the brain with axial sections showing multiple ring enhancing lesions (Nocardia abscesses) in the left occipito-parietal lobe and (B) showing increase in size of the abscesses with perilesional oedema causing midline shift to the right; (C) depicts frank pus drained from the largest abscess; and (D) depicts the large surgical scar mark corresponding to craniotomy. Microscopic findings (E, F) of drained pus showed modified acid-fast positive beaded filamentous branching hyphae suggestive of Nocardia farcinica confirmed by species subtyping. Repeat contrast-enhanced MRI of the brain with axial sections (G, H) show near-complete resolution of the occipito-parietal mycetoma following surgical evacuation and therapy with trimethoprim/ sulfamethoxazole and imipenem.**

# Epilepsy surgery in a patient with a diffuse EEG and a focal lesion

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We report an interesting case of a fifteen-year-old right handed male, with history of seizures since seven months of age, treated at the Fortis hospital, Mulund, Epilepsy Center. He had a history of perinatal insult due to hypoglycaemia and delayed milestones. Master (Vikas (name changed) had his first seizure which was with high grade fever. He remained seizure free until 2 years of life when he had episodes of generalized tonic posturing with loss of consciousness as his first seizure type. He was started on seizure medications. However, he continued to have frequent seizures despite two medications. He then developed violent head drops since the age of 4 years. These episodes were very frequent and occurred 10-15 times each day. He would injure himself and his mother used a helmet every time he stepped out of the house. The disabling nature of the episodes completely disrupted the life of this young kid. Even when he did not have the major episodes, he would remain in a sleepy state due to the subclinical (ELECTROGRAPHIC SEIZURES). He was being evaluated at several places and epilepsy surgery was written off as an option in view of his diffuse EEG changes. After several years of struggle, Vikas's parents reached our team at Fortis, Mulund. He was worked up for epilepsy surgery after a detailed video EEG, MRI, PET, neuropsychology evaluation. After detailed, discussion,

Dr Rima Chaudhari (consultant Epileptologist and neurologist in charge of the case) and her team decided to proceed with a resection of the right parieto occipital region. The child remains seizures free since then. At the last 3 months follow up his mother endorses a significant improvement in his overall cognition and behaviour and school performance. His EEG (electroencephalogram) has normalized for the first time in his life. (Figure 1)

**Figure 1: Playing cricket for the first time in his life**

# Looking Beyond Pain - Careful Clinical Examination Can Yield Surprising Results!!

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## Introduction

Localization in neurology is a challenging but the single-most important part of evaluation in any patient with neurological illness. The process of localization begins with the history which is then consolidated by examination and confirmed by radiological findings. Once this process is completed, then only patients can be diagnosed and treated with satisfactory results.

## Case Summary

We present a case of a 37-year-old female with history of pain in the right side of neck and right upper limb since two and a half years. She was on Pregabalin 150mg, Oxcarbazepine 600mg, Duloxetine 40mg for pain relief with a diagnosis of Fibromyalgia, anxiety, and myofascial pain syndrome. There was no associated weakness or numbness of the limb, no radiating shock like symptoms, no lower limb symptoms, or bladder bowel symptoms.

On examination, her higher mental functions and cranial nerve examination were normal. Tone in all four limbs was normal. Her right Abductor Pollicis Brevis (APB) muscle was wasted. Power testing showed weakness in the right APB and the right first dorsal interossei. Sensory examination showed sensory dulling in the medial aspect of the forearm. Reflexes were preserved and plantar was flexor. (Figure 1)

With this examination, the lesion could be localized to two sites in the neuraxis. It could either be a C8-T1 radiculopathy or a lower brachial plexopathy.

Nerve conduction study showed inelicitable Compound Motor Action Potential(CMAP) from the right APB and absent Sensory nerve action potential (SNAP) from the right medial antebrachial cutaneous (MABC) and reduced SNAP amplitudes from the right ulnar nerve.

In view of involvement of motor fibers in the median nerve and sensory fibers in ulnar and MABC distribution, lower plexus involvement with Neurogenic thoracic outlet syndrome was suspected.

X-ray chest showed no evidence of large cervical rib. MRI Brachial Plexus protocol showed thickened nerves and STIR hyperintensity in the lower and middle trunk of the brachial plexus. (Figure 2)

The etiological possibilities considered for lower plexus thickening considered were

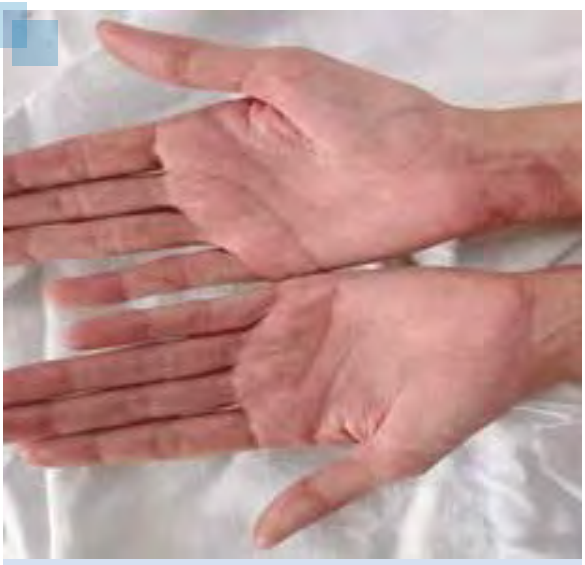

**Figure 1:**
**Right Abductor Pollicis Brevis wasting**

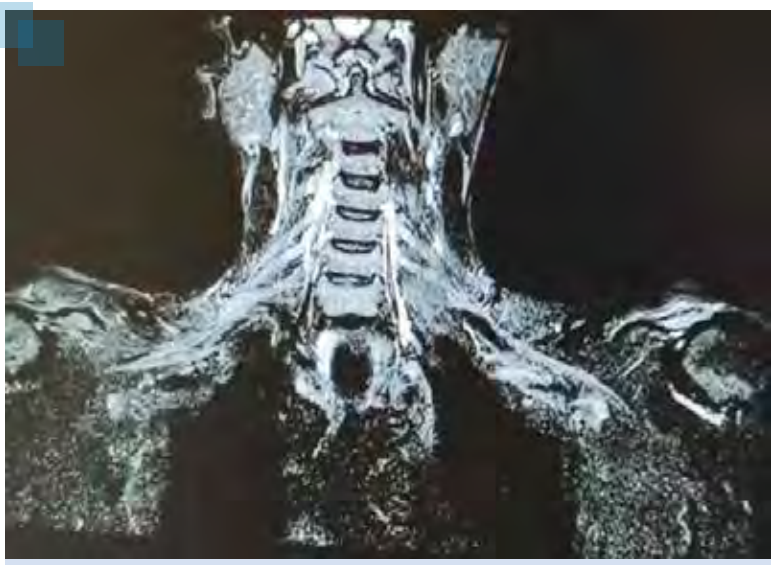

**Figure 2: Evident thickness and hyperintensity of trunk and cord of right brachial plexus**

1. Malignant infiltration- Neurolymphomatosis, carcinoma breast
2. Infective- Leprosy
3. Inflammatory- Sarcoidosis

Her blood work-up which included hemogram, liver and kidney function tests were normal. Peripheral blood smear and Serum LDH were normal. CSF analysis which included cytology, biochemistry, malignant cells was normal. Serum ACE levels were normal. As malignancy was considered, CT Chest & Abdomen, Mammogram, Ultrasound abdomen, tumour markers were done and all results were normal. PET scan was done which showed no FDG avid lesions.

She was counselled for fascicular biopsy and associated risk of weakness post the biopsy, which she denied. As infective causes were ruled out, she was started on intravenous pulse steroids for 5 days and then continued on 1mg/kg steroids. One-month post discharge she had significant relief in pain and now, does not require analgesics.

She has been planned for local exploration and biopsy from surrounding tissue if pain recurs on steroid taper.

## Conclusion

Clinical examination in this particular patient yielded an etiology of pain. The site of affliction and absence of weakness made biopsy taking in this patient risky in view of high possibility of a post-biopsy permanent deficit. Steroid administration resulted in resolution. Meticulous clinical examination is important in any patient with neurological illness

# Thunderclap Headache and Boomerang Sign in Dengue Encephalopathy

Source:- https://mansapublishers.com/index.php/ijcr/article/view/3096/2526

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An 18-year-old boy presented with high-grade fever with rashes over the body for 2–3 days followed by abrupt onset, severe holocranial headache associated with photophobia, phonophobia, nausea, and vomiting. Headache was so severe and sudden that the patient was immediately rushed to the emergency. The patient also developed slurring of speech and drowsiness later that day. On examination, the patient was febrile with a temperature of 101 F, his blood pressure was 100/60 mmHg, pulse rate was 110/min, and respiratory rate was 24/min. The patient was irritable, with irrelevant talking and restlessness. Cranial nerve, motor, sensory, and cerebellar examination were normal. His headache subsided with intravenous analgesics but fever and rashes persisted. Speech and sensorium improved along with headache. On investigation, his platelet counts were low (24,000/μL) and pyrexia workup showed dengue NS1 antigen positive. Brain magnetic resonance imaging (MRI) showed diffusion restriction in diffusion-weighted imaging images in the splenium of corpus callosum. (Figure 1)

Magnetic resonance angiography and venography were normal and no contrast enhancement was seen. The rest of the investigations were normal. The patient's fever, rashes, and platelet counts improved over a few days, and a repeat brain MRI after 2 weeks showed no diffusion restriction. (Figure 2)

Transient splenium hyperintensity in MRI is called as Boomerang sign due to its resemblance with boomerang [1]. It can be seen in infective encephalitis/encephalopathy, post-ictal state, anti-epileptic drugs withdrawal, toxic or metabolic encephalopathy, and occasionally in few primary headaches such as migraine and hemicrania continua [2-4]. In this case, probably dengue encephalopathy associated with capillary leakage caused neuronal damage in the splenium of the corpus callosum and hence may have triggered the cortical spreading depression causing headache.

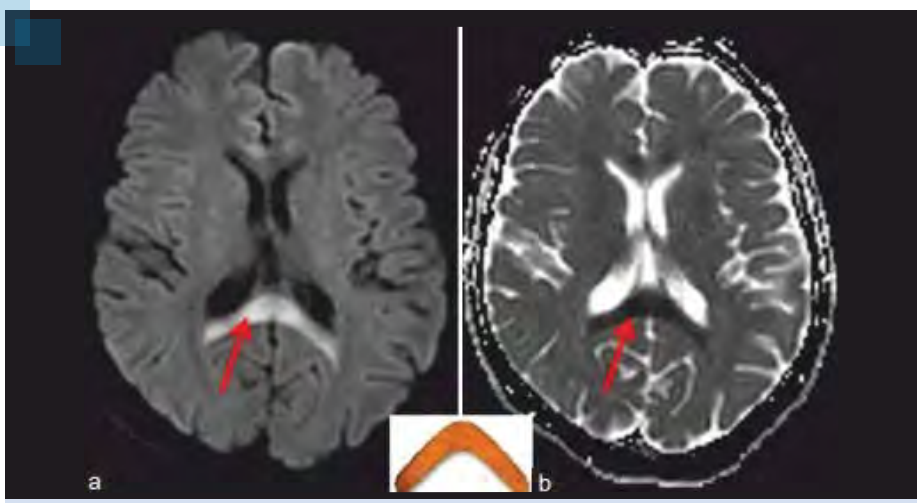


**Figure 1a and b: Diffusion-weighted imaging and apparent diffusion coefficient images showing diffusion restriction in splenium of corpus callosum (red arrows) at presentation (a and b). Inset image showing wooden boomerang, resembling restricted area in splenium of this patient**

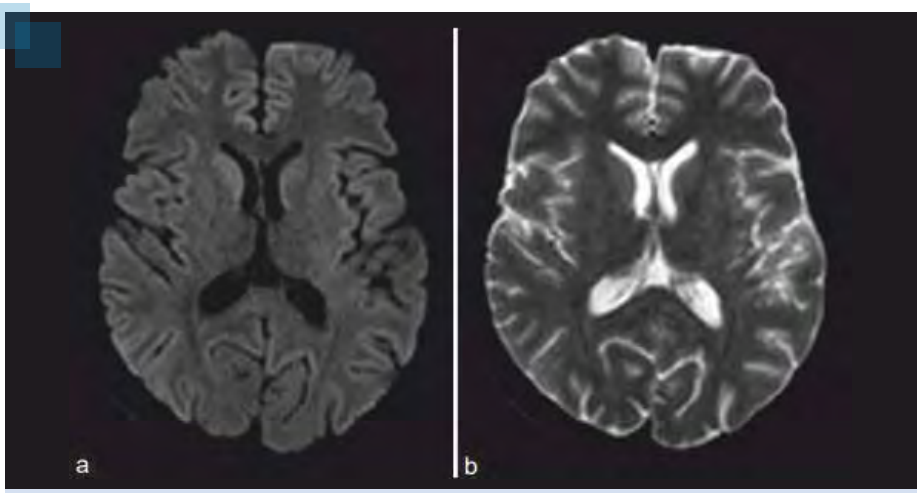


**Figure 2a and b: Diffusion-weighted imaging and apparent diffusion coefficient images showing disappearance of diffusion restriction from splenium of corpus callosum after 2 weeks (a and b)**