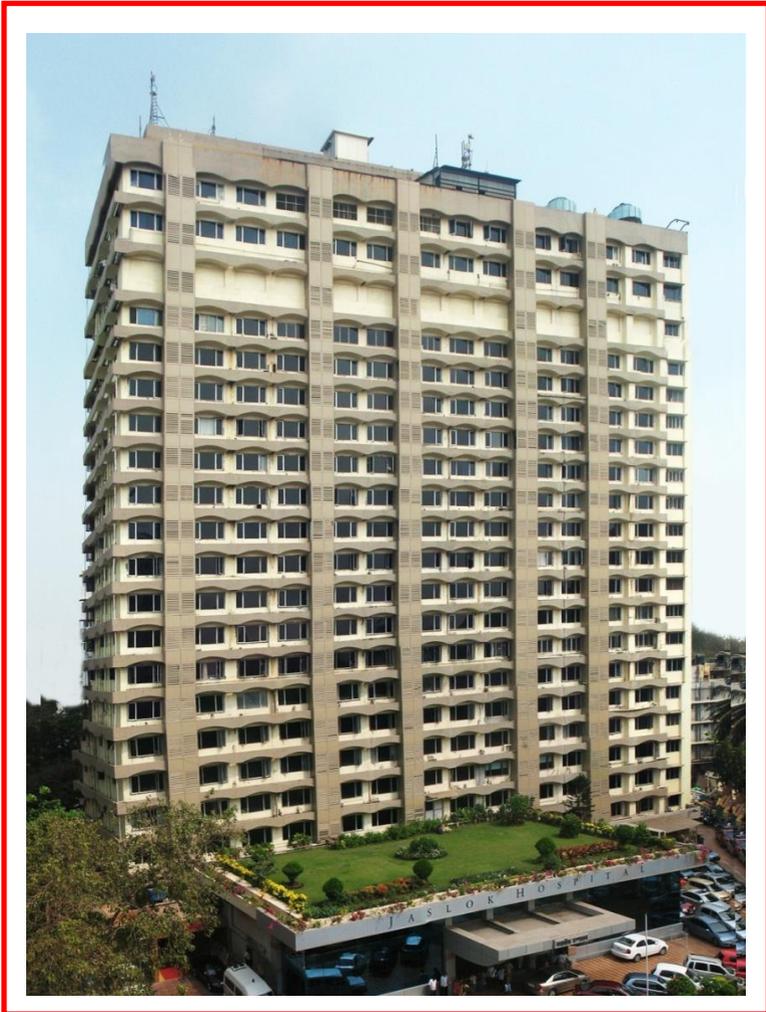




Jaslok Hospital & Research Centre, Mumbai

**Research eBulletin**

**Vol 2. Issue 4. Oct-Dec 2016**



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# Editorial

## **Putting our eBulletin together**

Over the past two years I have had the privilege of working with a group of remarkable individuals: our editorial team. Dr. Tarang Gianchandani despite her punishing schedule as CEO, reads every issue of the bulletin diligently and readily offers helpful suggestions that enhance its quality.

Dr. Fazal Nabi, our honorary paediatrician, in addition to his vital role in our Ethics Committee and the Apex Committee for Research, reviews every abstract to ensure our hospital's research gets a fair representation. Dr. Prochi Madon, our brilliant geneticist, helps in the selection of abstracts while microscopically proof-reading all the way through to printing. She sees that the abstracts are lined up one issue in advance to ensure timely release of the eBulletin.

Dr. Nihar Mehta, our honorary cardiologist, brims with ideas and suggestions. He is also quick in implementing them. The 'Legends of Jaslok' series is his brain child and he ensures that every issue has one of our legends featured as a source of inspiration to all. Dr. Pravin Agarwal, our honorary gastroenterologist, has helped design the bulletin in its first issue and continues to oversee its layout in addition to handling its research news. Ms. Maherra Khambaty, our editorial assistant, is involved in every stage of the eBulletin's production, especially in the quality control. Ms. Sabina Sawliwala ensures timely and quality printing. Ms. Viloo Williams and her team in the IT department email the bulletin to the consultants and others on time. Finally, Mr. George Alex and his marketing team get the bulletin up on the website and keep track of its readership and utilization. I am blessed to have such amazing colleagues. They make it possible for me to enjoy the entire process of bringing out this eBulletin.

In the 'Legends of Jalsok' series we feature eminent neurosurgeon Dr. H.N. Dastur who has been associated with our institution since its inception. I have been fortunate to interact with him on patient consultations. One is always impressed with his exquisite courtesy and grace. He carries an old world charm and compassion that is increasingly becoming rare these days. Dr. Sunil Pandya who deserves a legends piece in his own right has known him intimately over several years and so we requested him to write the tribute.

We look forward to receiving articles and suggestions for ensuring that the eBulletin retains academic interest among the Jaslok fraternity and in due time extends its reach well beyond the corridors of our hospital to garner a readership of medical professionals across the world.

**Rajesh M. Parikh, M.D., D.P.M., D.N.B.**  
**Director, Medical Research**

# Research News

The Indian Journal of Medical Ethics has been increasingly getting international recognition. Dr. Sunil Pandya, Hon. Neurosurgeon at Jaslok, has been the founder-editor of this prestigious journal and continues to be its editor emeritus. At a time when some of the most well-known international medical journals are facing increasing criticism over conflicts of interest or inappropriate vetting of authors and articles, the Indian Journal of Medical Ethics stands tall with its unblemished sterling record.

## Abstracts

### **Clinical features, treatment, and outcome of macrophage activation syndrome complicating systemic juvenile idiopathic arthritis: a multinational, multicenter study of 362 patients**

*Minoia F, Davì S, Horne A, Demirkaya E, Bovis F, Li C, Lehmborg K, Weitzman S, Insalaco A, Wouters C, Shenoi S, Espada G, Ozen S, Anton J, Khubchandani R, et al.*

**Arthritis and Rheumatology 2014;66:3160-9.**

**OBJECTIVE:** To describe the clinical, laboratory, and histopathologic features, current treatment, and outcome of patients with macrophage activation syndrome (MAS) complicating systemic juvenile idiopathic arthritis (JIA).

**METHODS:** In this multinational, multicenter study, pediatric rheumatologists and hemato-oncologists entered patient data collected retrospectively into a web-based database.

**RESULTS:** A total of 362 patients, 22% of whom had MAS at the onset of systemic JIA, were included in the study by 95 investigators from 33 countries. The most frequent clinical manifestations were fever (96%), hepatomegaly (70%), and splenomegaly (58%). Central nervous system dysfunction and hemorrhages were recorded in 35% and 20% of the patients, respectively. Platelet count and liver transaminase, ferritin, lactate dehydrogenase, triglyceride, and d-dimer levels were the sole laboratory biomarkers showing a percentage change of >50% between the pre-MAS visit and MAS onset. Evidence of macrophage hemophagocytosis was found in 60% of the patients who underwent bone marrow aspiration. MAS occurred most frequently in the setting of active underlying disease, in the absence of a specific trigger. Nearly all patients were given corticosteroids, and 61% received cyclosporine. Biologic medications and etoposide were given to 15% and 12% of the patients, respectively. Approximately one-third of the patients required admission to the intensive care unit (ICU), and the mortality rate was 8%.

**CONCLUSION:** This study provides information on the clinical spectrum and current management of systemic JIA-associated MAS through the analysis of a very large patient sample. MAS remains a serious condition, as a sizeable proportion of patients required admission to the ICU or died.

## **Oncotic Cerebral Aneurysms in a Case of Left Atrial Myxoma, Role of Imaging in Diagnostics and Treatment**

*Gupta MM, Agrawal N*

**Polish Journal of Radiology 2015;80:490-5.**

**BACKGROUND:** Myxomatous cerebral (oncotic) aneurysms following atrial myxoma is a rare neurological complication.

**CASE REPORT:** We report an 11-year-old boy with left atrial myxoma and multiple cerebral oncotic aneurysms. The characteristics of these aneurysms are indefinite and variable. The "Metastasize and Infiltrate" theory may be the key mechanism in the formation of these aneurysms.

**CONCLUSIONS:** Magnetic resonance imaging (MRI), computed tomography (CT) and angiography are useful in the diagnostics while digital subtraction angiography (DSA) is the best option. There are no definite guidelines for therapy of these aneurysms. Resection of cardiac myxomas, chemotherapy, radiotherapy, coil embolization and surgical treatment could be helpful.

## **Coexistence of atypical hemolytic uremic syndrome with membranoproliferative glomerulonephritis and antineutrophil cytoplasmic antibodies-associated vasculitis**

*Sathe KP, Mehta KP*

**Saudi Journal of Kidney Diseases and Transplantation 2016;27:800-4.**

The simultaneous presence of multiple immune-mediated diseases in a single host is rare. The implications of such coexistence relating to the disease pathogenesis and treatment are not well understood. We describe two cases of renal failure with immune-mediated overlap conditions. We believe, this is the first reported case of antineutrophil cytoplasmic antibody (ANCA) associated vasculitis coexisting with atypical hemolytic uremic syndrome (HUS). Two boys aged eight years (Case 1) and 10 years (Case 2) presenting with renal failure secondary to glomerulonephritis are described. Based on the clinical features, a detailed immunological workup and kidney biopsy was performed to arrive at the diagnosis. Immune-mediated renal dysfunction was present in both cases. Screening for other coexisting autoimmune phenomenon was performed based on suspicious clinical features. Case 1 presented with renal failure and D-HUS with low serum C3. Renal biopsy revealed membranoproliferative glomerulonephritis Type 1. The child improved following treatment with plasma infusions and steroids. Case 2 presented with ANCA-positive vasculitis. Renal biopsy was suggestive of focal mesangioproliferative glomerulonephritis. Disease course was further complicated by D-HUS with low serum C3. Factor H antibody was positive. Complete renal recovery was documented following treatment with intravenous rituximab, steroids, cyclophosphamide, and plasmapheresis. Screening for the presence of coexisting autoimmune diseases is imperative to identify covert immune-mediated pathologies and for the successful overall management of such cases.

## **Vitamin D as a marker of cognitive decline in elderly Indian population**

*Tejal Kanhaiya Vedak, Vaishali Ganwir, Arun B. Shah, Charles Pinto, Vikram R. Lele, Alka Subramanyam, Hina Shah, and Sudha Shrikant Deo*

**Annals of the Indian Academy of Neurology 2015;18:314-9.**

**Objectives:** Very few studies in India have addressed the role of vitamin D in cognitive function. The present study was conducted to assess the serum levels of 25-hydroxyvitamin D (25(OH)D) and its association with markers of cognitive impairment and homocysteine levels in the elderly Indian population.

**Materials and Methods:** The study population consisted of patients with dementia (Group A,  $n = 32$ ), mild cognitive impairment (MCI; Group B,  $n = 24$ ), and elderly age-matched controls (Group C,  $n = 30$ ). Measurement of serum levels of 25(OH)D and total homocysteine were done.

**Results:** Significant decreased concentration of 25(OH)D and increased concentration of homocysteine was observed. Association of serum levels of vitamin D with markers of cognitive decline as well as serum homocysteine levels was observed in patients with dementia and MCI when compared to controls.

**Conclusion:** Correlation of vitamin D with markers of cognitive decline and homocysteine opens a new door for early diagnosis of cognitive impairment.

## **LACE versus BEAM conditioning in relapsed and refractory lymphoma transplant: retrospective multicenter analysis of toxicity and efficacy**

*Khattry N, Gupta A, Jain R, Gore A, Thippeswamy R, Jeevangi N, Kannan S, Nair R, Saikia T*

**International Journal of Hematology 2016;103:292-8.**

We compared the lomustine, cytarabine, cyclophosphamide and etoposide (LACE) and BCNU, etoposide, cytarabine, melphalan (BEAM) conditioning regimens for toxicity, engraftment kinetics, and efficacy in 139 patients undergoing autologous hematopoietic stem cell transplant for primary refractory or relapsed lymphoma. Ninety-two patients with Hodgkin lymphoma and 47 with non-Hodgkin lymphoma were enrolled. Seventy-five patients received LACE while 64 received BEAM. The incidence of grade 3-4 oral mucositis (9 vs 38%;  $P < 0.001$ ) and parenteral nutrition requirement (32 vs 69%;  $P < 0.001$ ) were significantly lower in the LACE cohort. The median days to myeloid (10 vs 11;  $P = 0.007$ ) and platelet engraftment (13 vs 15;  $P = 0.026$ ) were shorter for the LACE cohort. Transplant-related mortality in the LACE group was 9% compared to 13% in patients treated with BEAM ( $P = \text{NS}$ ). The probability of overall survival (OS) and progression-free survival (PFS) at 5 years for entire cohort was 46 and 41%, respectively. Probability of OS (LACE 46% vs BEAM 47%;  $P = \text{NS}$ ) and PFS (LACE 37% vs BEAM 47%;  $P = \text{NS}$ ) at 5 years was comparable between two groups. We conclude that LACE has better toxicity profile compared to BEAM and results in similar long-term survival in primary refractory or relapsed lymphoma transplant.

## **Cerebral aneurysm treatment in India: Results of a national survey regarding practice patterns in India**

*Ambekar S, Madhugiri V, Pandey P, Yavagal DR*

**Neurology India 2016;64 Suppl:S62-9.**

**BACKGROUND:** The management of intracranial aneurysms (IAs) varies widely depending upon a number of factors.

**OBJECTIVE:** To understand the variations in practice patterns in the treatment of IAs in India.

**METHODS:** The survey consisted of 23 questions. Two group emails were sent to members of the Neurological Society of India and the Neurological Surgeons Society of India. Uni- and multivariate analysis was performed where appropriate.

**RESULTS:** The response rate was 10.13% (150/1480). Fifty percent of the respondents used steroids in subarachnoid hemorrhage and 64% initiated triple-H therapy prophylactically. There was no significant difference in the use of steroids, antifibrinolytics, mannitol, or hypertonic saline and the choice of therapeutic intervention (clipping or endovascular therapy [EVT]) for anterior circulation aneurysms between physicians working at teaching and nonteaching hospitals. However, physicians in teaching and government hospitals were less likely to choose EVT for middle cerebral artery aneurysms as the first line of treatment (odds ratio [OR] 0.6 and 0.1, respectively). Physicians working at private hospitals were more likely to have EVT facilities than those working in government-owned hospitals. On multivariate analysis, physicians working in teaching hospitals preferred surgical clipping to EVT for posterior circulation aneurysms (OR = 0.7) and physicians at teaching hospitals performed >50 cases/year.

**CONCLUSION:** Our study demonstrates the prevailing practice patterns in the management of IAs in India. Surgical clipping is the preferred treatment of choice for anterior circulation aneurysms and EVT for aneurysms along the posterior circulation. Corticosteroids and prophylactic "triple-H" therapy are still used by a large proportion of physicians.

## **Legends of Jaslok**



**Dr. Homi M. Dastur** has been associated with Jaslok Hospital and Research Centre since its inception. He set up the practice of neurosciences at a very high standard together with Dr. Gajendra Sinh and Dr. Noshir Wadia. The latter two doyens are unfortunately no more with us. Stereotactic surgery, surgery of intracranial aneurysms and arteriovenous malformations was started at Jaslok, in addition to the more common neurosurgical operations. He had an enviable expertise in neuroradiology, having worked with Dr. James Bull, one of the pioneers of percutaneous carotid angiography. Dr. Srinivas Desai was encouraged to develop interventional radiology. All patients, rich and poor, were treated with the same attention and care.

Dr. Dastur was born in 1926. He returned to his *alma mater*, Seth G. S. Medical College and King Edward VII Hospital, forsaking a promising career in the Department of Neurosurgery at Atkinson Morley's Hospital, London, at the behest of his *guru*, Dr. A. V. Baliga, to set up a department of neurosurgery. Starting with minimal facilities, he painstakingly set about his task. He broke several traditions. He insisted on having daily sessions in the out-patient department even though this meant increasing his own workload. When administrative protests met this proposal, he showed how many of his patients travelled long distances, at times hundreds of kilometres, to come to the hospital. When they did reach it, they were often in precarious states with severely raised intracranial pressure or severe compression of the spinal cord. If made to wait till the next weekly outpatient clinic, they would either die or be rendered paraplegic. His request was granted.

When he left the department, Dr. Dastur had also created a unique system of medical records, a library of books and journals and an ethos of caring sincerely for the poorest of patients. Every doctor referring a patient to the department was sent a detailed reply when the patient was ready to go home, describing the clinical diagnosis, findings on tests and at surgery and recommending further treatment after the patient returned to his doctor.

Clinical and clinico-pathological studies on tuberculosis of the central nervous system, craniovertebral anomalies and other conditions were published by him in national and international journals and are oft-quoted even today.

Dr. Dastur is enjoying a well-deserved retirement with his family and friends. He continues to indulge in his study of birds, flowers (especially orchids) and enjoy Western classical music. May he continue to do so for long.

## Research Events

### Workshops held

- a) 'The Next Step: From Dissertation to Publication' by Dr. Nihar Mehta on 12/09/2016.
- b) 'Evidence Based Medicine' by Dr. Nihar Mehta on 19/09/2016 for nursing staff.
- c) 'Research Statistics' by Dr. Maninder Singh Setia on 20/09/2016 for DNB students.
- d) 'Research Methodology' by Dr. Pravin Agrawal on 27/09/16 for nursing staff.

### Editorial Board

Drs. Tarang Gianchandani, Rajesh Parikh, Fazal Nabi, Nihar Mehta, Prochi Madon & Pravin Agrawal.

Coordinating Editors: Drs. Prochi Madon & Nihar Mehta.

Editorial Assistant: Ms. Maherra Khambaty.