

**R65 MAJOR ADVERSE CLINICAL EVENTS (MACE) IN THOSE WITH UPPER EXTREMITY ATHEROSCLEROTIC DISEASE**

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**Objectives:** Compare major adverse clinical events (MACE) between patients with upper extremity arterial disease to those with lower extremity arterial disease.

**Background:** Upper extremity peripheral arterial disease, although occurring less frequently than lower extremity peripheral arterial disease, negatively impacts one's lifestyle. Most of the studies on peripheral arterial disease have focused on the lower extremities and the clinical impact of upper extremity arterial obstructions remains less well known. We postulate that patients with upper extremity arterial obstruction have similar major adverse clinical event rates as those with lower extremity arterial disease.

**Methods:** 67 consecutive patients with upper extremity arterial studies and 370 consecutive patients with lower extremity arterial studies performed in 2004 were followed for at least two years. MACE such as death, stroke, transient ischemic attack, myocardial infarction and unstable angina for at least a two-year period were tabulated and compared between the two groups. Additional evaluations also were made between those with and without upper extremity arterial disease. The final result was adjusted for diabetes mellitus, hypertension, hyperlipidemia, and renal insufficiency. One-tailed t test was used.

**Results:** A total of eight (28%) events occurred in 29 patients with abnormal upper extremity arterial study and 24 (21%) events in 112 patients with abnormal lower extremity arterial study, p=0.28. Between those with normal and abnormal upper extremity arterial study, 38 patients with normal upper extremity arterial study had a cumulative event rate of five (13%) versus 28% in those with abnormal upper extremity arterial study, p=0.02. Comparing MACE in those with abnormal lower extremity arterial study, 20 events (7.75%) occurred in patients with normal lower extremity arterial study, p<0.0001.

**C1 NOT ONLY SKIN DEEP: DISABLING PANSCLEROTIC MORPHEA**

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**Introduction:** Disabling pansclerotic morphea, which characteristically spares the fingers and toes, is the rarest, most severe form of localized scleroderma.

**Case Report:** An 18-year-old Hispanic male with a history of morphea presented with a foot ulcer. His disease began insidiously at the age of 10 with small, bilateral lower extremity plaques, which progressed upward. On physical exam, the trunk, back, face, neck, proximal upper extremities, thighs, legs, and dorsum of the feet were covered with confluent areas of sclerotic hypopigmentation intermixed with patchy areas of hyperpigmentation. His toes, soles of the feet, distal upper extremities, and scalp were largely spared. There was a 5 x 6 cm ulceration on the dorsum of the left foot. Decreased range of motion in the ankles and subtalar joints, and moderate muscle wasting of the lower extremities were noted. Laboratory tests revealed a sedimentation rate of 65 and CRP level of 32.0. ANA profile was negative. A skin biopsy at an outside facility confirmed the diagnosis of morphea. The patient received methotrexate, corticosteroids, IV antibiotics, and wound care.

**Discussion:** This case illustrates the rarest form of systemic sclerosis and its typical features, including generalized sclerotic skin lesions with sparing of the toes and fingers (in this case near complete distal upper extremities sparing). Characteristically, in disabling pansclerotic morphea there is involvement of deep tissues and other structures, including muscle and bone. This patient's decreased active range of motion suggests that muscle involvement is progressing. Limited references note the benefits of corticosteroid treatment combined with low dose methotrexate.

**C2 MASSIVE OSTEOLYSIS DUE TO FAILURE OF A UNIPOLAR ENDOPROSTHESIS: TWO CASE REPORTS**

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**Background:** These cases report the failure in the polyethylene component used within an Osteonics unipolar endoprosthesis.

**Case Reports:** A 69-year-old woman with a left displaced femoral neck fracture was treated using an Osteonics unipolar endoprosthesis. This design has a polyethylene core in the head which was not intended for motion. She required a revision to a total hip arthroplasty. Her pre-revision radiographs showed a crack through the endoprosthesis coupling between the head and stem, with erosions into the rami, ischium and the medial femoral cortex. The second case, a 55-year-old female with a right displaced femoral neck fracture, was also treated with an Osteonics unipolar endoprosthesis. She had a revision, 9½ years later, to a tumor prosthesis with an articulating bipolar prosthesis. This patient also had massive osteolysis of the proximal femur and acetabulum. In both of these cases, failure of the prosthesis led to the core being dislodged from the metal cap, and created an articulating surface. The polyethylene had large areas that had basically been carved out from direct wear. This caused a large amount of polyethylene debris, which we believe was responsible for this massive osteolysis.

**Discussion:** We report these cases to heighten the awareness of a polyethylene component in this particular Osteonics endoprosthesis head, and we question the need for implanting polyethylene in any prosthetic in which it is not being directly used for the bearing surface. We believe these particular types of failures would be significantly decreased in a comparable prosthesis with a purely metallic head.

**C3 ACUTE DRUG-INDUCED HEPATITIS ASSOCIATED WITH NONI JUICE INGESTION**

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**Introduction:** The use of complementary and alternative medicine is growing rapidly. Unfortunately, many patients are reluctant to include herbal remedies among their listed medications.

**Case Report:** An 80-year-old female had generalized pruritus and jaundice for four days. She denied gastrointestinal symptoms or fever. She also denied the use of prescriptions, illicit drugs, alcohol, or over-the-counter medications. The physical exam revealed scleral icterus and diffuse jaundice with excoriations. She had no stigmata of chronic liver disease. Results of her abdominal exam were benign. Laboratory values were significant for: total bilirubin 13.6 mg/dL, direct bilirubin 9.7 mg/dL, AST 87 U/L, ALT 82 U/L, and alkaline phosphate 279 U/L. Abdominal ultrasound with Doppler revealed only cholelithiasis. Viral hepatitis panel was negative. Antinuclear antibody and antimitochondrial antibody were negative. Antismooth muscle antibody titer was 1:80. Immunoglobulin levels were normal. A liver biopsy showed portal lymphocytic infiltrates with occasional plasma cells and eosinophils and steatohepatitis. Drug-induced hepatitis was suspected. Upon further questioning, the patient admitted to drinking Noni juice daily for one month. Four cases of acute hepatitis related to Noni juice have been reported previously.

**Discussion:** Noni, a fruit of the *Morinda citrifolia* tree, has been ingested for centuries in Southeast Asia and the Pacific, where it is used as a remedy for many conditions. Recently, concentrated Noni juice has been aggressively marketed to Western

**C4 CHURG-STRAUSS-SYNDROME ASSOCIATED WITH LEUKOTRIENE RECEPTOR ANTAGONISTS (LTRA)**

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**Case Reports:** Churg-Strauss syndrome (CSS) is a rare vasculitic disorder that generally occurs in patients with bronchial asthma. CSS is being increasingly recognized in asthmatic patients treated with leukotriene receptor antagonists. The nature, however, of this relationship remains to be elucidated. The present report describes 3 asthmatic patients who developed clinical manifestations highly suggestive of CSS, although one patient lacked the presence of eosinophilia. That patient, however, exhibited biopsy-proven cutaneous necrotizing vasculitis, which improved following withdrawal of the leukotriene receptor antagonist (montelukast). The second patient presented with systemic constitutional signs including fever, malaise, arthralgias, clinical jaundice, peripheral blood eosinophilia, and biopsy-proven eosinophilic hepatitis. The third patient also had circulating eosinophilia, scleritis, and arthritis. All patients improved following discontinuation of montelukast.

The pathogenesis of LTRA-related CSS is not well understood. There are several hypotheses: 1) CSS develops primarily in asthma patients who had been taking LTRA, for an underlying eosinophilic disorder that was being masked by corticosteroid treatment and unmasked by novel asthma medication-mediated corticosteroid withdrawal. 2) Temporal relationship between the use of LTRA and the development of CSS (the natural progression of an underlying disease). 3) Leukotriene imbalance resulting from leukotriene receptor inhibition. LTRAs block the synthesis of LTC<sub>4</sub>, LTD<sub>4</sub>, and LTE<sub>4</sub>, but do not inhibit LTB<sub>4</sub> (a potent chemoattractant for eosinophils and neutrophils), and the modulation of adhesion molecules expression such as P-selectin. Leukotriene B<sub>4</sub> (LTB<sub>4</sub>), a chemotaxin without bronchoconstrictor activity, is the major member of the dihydroxy leukotrienes, and it has an entirely different role in the inflammatory process than the cysteinyl leukotrienes LTC<sub>4</sub>, LTD<sub>4</sub>, and LTE<sub>4</sub>. It has pleiotropic effects on different cell systems, and on monocytes and macrophages LTB<sub>4</sub> increases the production of IL-6, IL-1 and TNF-alpha, as well as chemotaxis and aggregation. LTB<sub>4</sub> increases T lymphocytes chemokinesis, chemotaxis, aggregation, adherence, activation, granule release, and superoxide production. LTB<sub>4</sub> also enhances B cell activation, differentiation, proliferation, immunoglobulin production, and NK cell activity. On the other hand, leukocytes from asthmatic patients produce large amounts of LTB<sub>4</sub>, which coupled to the high circulating levels of IL-5 in CSS (a potent stimulant of LTB<sub>4</sub>) may alter the balance in favor of LTB<sub>4</sub> over LTC<sub>4</sub>/LTD<sub>4</sub>/LTE<sub>4</sub> and lead to the accumulation of eosinophils, neutrophils, T and B lymphocytes, and monocytes/macrophages, which leads to a burst of the inflammatory process, and finally results in the clinical expression of CSS.

**C5 INFLUENCE OF DR. JOHN ADRIANI ON THE AMERICAN BOARD OF ANESTHESIOLOGY ORAL EXAMINATION**

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**Case Report:** Dr. John Adriani moved to New Orleans in 1941 to become the director of Anesthesiology at Charity Hospital. His incredible career would span more than three decades until his retirement in 1976. During this period, he was a world-renowned educator, clinician and author, whose prolific writings are still relevant in anesthesiology today. For his dedication and contributions, he was awarded the American Society of Anesthesiologists Distinguished Service Award in 1949. Less well known is Dr. Adriani's influence on the American Board of Anesthesiology (ABA) Oral Examination process. Dr. Adriani was an ABA Director from 1960-1972 and chaired the examinations committee from 1964 to 1965. During the time of his chairmanship, Dr. Adriani wrote a document entitled "The Oral Examination of the American Board of Anesthesiology," which was intended as a guide for examiners to help improve their examination skills. Dr. Adriani's paper focused on 3 areas: The criteria of a good anesthesiologist, the requisites of a good examiner, and the type of questions to be asked. Though this document has not been widely circulated, close study reveals the influence to this day of Dr. Adriani's beliefs about what constitutes a valid exam on the ABA Oral Examination process.

**C6 SARCOMA BOTRYOIDES OF THE UTERINE CERVIX IN A 48-YEAR-OLD FEMALE: A CASE OF MISTAKEN IDENTITY?**

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**Background:** Sarcoma botryoides, a variant of embryonal rhabdomyosarcoma, is a rare gynecologic malignancy, characterized by a typical grape-like appearance due to a layer of spindle cells pushing up beneath the mucosa in polypoid masses. It is most familiar as a tumor of the vagina of infants, but has been found in the cervix during adolescence and the reproductive years, or in the corpus uteri during the postmenopausal years.

**Case Report:** A 48-year-old female presenting with vaginal bleeding and cramping was initially diagnosed with recurrent cervical polyps. Dilatation and curettage with hysteroscopy revealed an enlarged uterus, multiple exophytic cervical polyps emanating from a single large stalk, and multiple endometrial polyps. Complete removal was difficult due to the large mass of the lesion. Initial diagnosis was malignant spindle cell tumor with cartilaginous islands, which was consistent with sarcoma botryoides of the uterine cervix. Following review of the pathology report and consultation with the patient, she underwent a total abdominal hysterectomy and bilateral salpingo-oophorectomy, which revealed a normal-sized uterus and cervix, normal ovaries and fallopian tubes, and no evidence of intra-abdominal metastatic disease. Final pathologic diagnosis was spindle cell tumor with cartilaginous island consistent with botryoid rhabdomyosarcoma. The specimens were sent to Johns Hopkins Hospital for consultation.

**Conclusions:** Cervical rhabdomyosarcoma in patients over 40 years of age is rare. A PubMed search for the terms cervical sarcoma botryoides and sarcoma botryoides of the uterine cervix reveals a total of 115 documented cases from 1867 through 2004. Since 1980, 58 cases have been reported, 5 of which occurred in women aged 40 and above. The differential diagnosis includes benign cervical polyps, adenocarcinoma, and pseudosarcoma botryoides. The final interpretation and diagnosis reported by Johns Hopkins Hospital was an atypical form of adenocarcinoma with rhabdomyoblastic differentiation and heterologous elements (cartilage), involving the cervix and lower uterine segment.

**C7 MAPPING THE MEDICATION ADMINISTRATION PROCESS VIA A MEDICAL BUSINESS PROCESS MODELING TOOL TO IDENTIFY OPPORTUNITIES FOR IMPROVEMENT**

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**Objective:** To document and visually map the medication administration process using a newly created Medical Business Process Modeling (medBPM) tool, specifically designed for use in healthcare. Mapping will include the following areas: 1. Order prescription and transcription; 2. Medication, medication related supplies and information gathering; 3. Medication preparation; 4. Medication administration; 5. Medication documentation; 6. Patient monitoring. Visual mapping of the medication administration process will provide valuable insight regarding the efficiency, safety, and status of regulatory and accreditation compliance throughout the process and will be used to determine process improvement opportunities for future projects.

**Case Report:** Traditional mapping methodologies do not adequately capture the family of resources, points of interaction between individuals, work practices, and information that make up complex healthcare processes. medBPM is a tool and a flow-charting methodology specifically designed to capture, analyze and optimize the business processes of the healthcare field. It combines the power of computerization for quantitative metric tracking, analytics and reporting and the expertise of people for qualitative process evaluation and reporting. This project was conducted according to the medBPM methodology, which involved on-site observations, shadowing of personnel, modeling, validation and subsequent off-site modeling, validation and analysis. The on-site process consisted of semi-structured interviews and observations with photos and video taping of the process.

**Results:** medBPM workflow models included: 1. Pharmacy workflow for first doses and STAT doses; 2. Pharmacy workflow: Refill/scheduled doses; 3. Nursing medication administration process: Critical care unit; 4. Nursing medication administration process: Medical surgical unit; 5. A macro model showing the overall medication ordering, preparation, administration and documentation process. Initial identification of process improvement provided opportunities for nursing and pharmacy related to the medication administration process based on workflow models.

**Conclusions:** Multi-disciplinary work groups were utilized to develop action plans for identified process improvement opportunities. Rapid cycle targets included: 1. To standardize two patient identifiers; 2. Standardize reference to medication information for medication administration; 3. To reduce amount of medication returns; 4. To modify Pyxis refill time.

C8 IMPLEMENTATION OF CODE GREEN

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**Case Report:** Following Hurricane Katrina, Ochsner experienced an influx of patients who presented unique challenges. Patients formerly managed by the local charity system were processed instead through our Emergency Department, increasing our uninsured population from 4% to 11%. Concurrent with this demographic shift, our Emergency Department and hospital witnessed an upsurge in patients and families who displayed agitated and/or combative behavior. The community suffered from a decrease in available nurses, security personnel, and virtually all other categories of workers. The staff, including the Hospital Medicine staff that managed 60% of the inpatient population, was challenged to find effective ways to deal with disruptive behaviors displayed by patients and by their families. A new code, Code Green, was devised. When a Code Green is called over the overhead paging system, all available personnel who have been trained in crisis intervention proceed quickly to the area of the code to help diffuse the situation. They respond to threats of a physical, verbal or non-verbal nature. Only anecdotal outcome data are available at present. Patient Care Coordinators have reported that patients who are agitated respond very positively to the arrival of several male personnel who interact with the patient in a calming manner. Widespread training of staff in crisis management techniques, as well as initiating a special code for assistance with combative patients helped our organization cope with a sudden shift in patient demographics following a natural disaster in our area. The innovative efforts of the organization helped retain staff during a particularly challenging time.

C10 PARATHYROID HORMONE ANALOG TARGETED TO BONE IS MORE EFFECTIVE AT INCREASING BONE MINERAL DENSITY IN MICE

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**Case Report:** Osteoporosis is a major health issue affecting approximately 10 million Americans. Bisphosphonate compounds are currently the main treatment for osteoporosis; these compounds contain a pyrophosphate group which targets these compounds to the bone mineral matrix, where they act by reducing bone resorption. Targeting the drug to its intended site of action reduces systemic side effects and extends the duration of action of the drug to yearly parenteral dosing. Parathyroid hormone (PTH) is a powerful anabolic agent in bone and provides superior efficacy in the treatment of osteoporosis. However, unlike bisphosphonates, this agent must be injected daily and can cause undesirable side effects, including hypercalcemia. To target PTH directly to bone similarly to bisphosphonates, we have synthesized two PTH analogs which contain the biologically active portion of the PTH molecule (amino acids 1-34) linked to a collagen-binding domain that binds tightly to the bone's collagen matrix. We have shown previously that these PTH analogs bind type 1 collagen with similar affinity to that of the collagen binding domain alone, and they activate cAMP accumulation in cells transfected with the PTH receptor with similar potency and efficacy to that of PTH[1-34]. When injected into female mice (weekly dosing), the analogs cause increases in bone mineral density which exceed those seen after weekly therapy with PTH alone. In vivo efficacy with longer (i.e., monthly) dosing intervals is currently being assessed. Compounds such as these may ultimately allow the superior anabolic effect of parathyroid hormone to be obtained with the dosing convenience of the bisphosphonate compounds.

C9 NOVEL COL1A2 MUTATION LEADING TO OSTEOGENESIS IMPERFECTA TYPE 1 IN IDENTICAL TWINS C11 EFFECTS OF STRESS AFTER HURRICANES KATRINA AND RITA ON PUBERTAL DISORDERS IN CHILDREN

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**Case Report:** Osteogenesis imperfecta (OI) is an autosomal dominant disorder of bone collagen leading to multiple fractures. It is classified based on severity, with type 1 being the mildest. Most individuals with OI have mutations in one of two genes encoding type 1 collagen, COL1A1 or COL1A2. We now report on identical twins with OI type 1 from a novel COL1A2 mutation. By age 16, twin 1 suffered 5 fractures, and twin 2 suffered 3 fractures. Each twin had increased bone turnover (elevated alkaline phosphatase and urinary crosslinked N-telopeptides). The spinal bone mineral density of each twin was low (twin 1 Z=-2.6, twin 2 Z=-2.3). Their mother sustained one fracture and had been diagnosed with osteopenia, treated with risenedronate and alendronate. Sequence analysis for each twin revealed a C3495>G missense mutation in COL1A2. No other mutations were found in COL1A1 or COL1A2. Sequence analysis of the parents revealed the same mutation in the mother but not in the father. This mutation is predicted to cause a D1165E change in alpha-2(I) collagen. This substitution results in a conservative amino acid substitution in the C-propeptide region of the protein. Alterations of Asp1165 have not been previously described, either in OI or as a known polymorphism. There are few mutations in the C-propeptide region of alpha-2(I) collagen, and most of those cause more severe disease. We postulate that although the mutation occurs in an apparently critical region, the substitution is conservative and, thus, the bone fragility in this family is relatively mild.

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**Case Report:** The magnitude and scope of Hurricanes Katrina and Rita are such that they affected the entire referral base for the pediatric endocrine practice at Ochsner Medical Center. Essentially, all families in this region were affected by these events, either directly or indirectly. This resulted in increased stress levels for both parents and children. Several studies provide evidence for effects of stress on pubertal development; some found that stress induces puberty, while others showed that stress postpones puberty. We examined the effects of the stress caused by Hurricanes Katrina and Rita on pubertal development in our patient population by conducting a retrospective chart review; the incidence of precocious and delayed puberty was compared over a 4.5 month period preceding and following the storms. The total number of new patients referred and the incidence of diagnoses that are unaffected by stress (i.e., thyroid disease) were essentially unchanged. On the other hand, the incidence of precocious puberty decreased by 52% after the storm, while the incidence of pubertal delay increased by 9% in the post-storm period. This study thus provides evidence that stress delays the onset of puberty in children.

**C12 ORBITAL PSEUDOTUMOR IN A PATIENT WITH SERONEGATIVE RHEUMATOID ARTHRITIS**

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**Introduction:** Orbital pseudotumor is a rare ophthalmologic manifestation of autoimmune disease. It is a condition that has an autoimmune origin. This condition is associated with rheumatoid arthritis but also can be associated with other autoimmune diseases such as systemic lupus erythematosus and Wegener's granulomatosis.

**Case Report:** We present a case of a 35-year-old female with a two-week history of intermittent right eyelid erythema, swelling, and ptosis. She also had blurred vision for the same amount of time. She has a history of seronegative rheumatoid arthritis which was diagnosed four years prior to presentation. MRI showed a soft tissue mass present in the superior aspect of the right orbit. A biopsy done later was consistent with inflammatory pseudotumor. She was given prednisone 60mg daily for her symptoms. After one week her symptoms had improved.

**Discussion:** Orbital pseudotumor is an inflammatory disorder that has an autoimmune origin. It is associated with autoimmune disorders such as Wegener's granulomatosis, Churg-Strauss syndrome, systemic lupus erythematosus, and rheumatoid arthritis. Orbital MRI is an important diagnosis tool. Biopsy is usually not done unless the patient doesn't respond to steroid treatment. Steroids are usually the initial treatment. Radiotherapy or chemotherapeutic agents such as cyclophosphamide, methotrexate, or cyclosporine are used if there is no response to steroids.

**C14 EOSINOPHILIC FASCIITIS INDUCED BY FIRE ANT BITES**

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**Introduction:** Eosinophilic fasciitis is a rare scleroderma-like disorder first described by Shulman in 1974. Eosinophilic fasciitis may be triggered by vigorous exercise, drugs, borreliosis, arthropod bites, and trauma. We report a case of eosinophilic fasciitis, confirmed by biopsy, secondary to fire ant bites.

**Case Report:** A 79-year-old female was in her backyard cleaning up debris after Hurricane Katrina barefooted and had numerous fire ant bites. One week later she noted swelling of her right leg and underwent further evaluation, including a venous Doppler which was negative for a thrombus. The swelling in her right leg was associated with the skin being thickened, indurated and bound down. She was seen by a dermatologist, who performed a punch biopsy of the skin which revealed dermal edema with chronic inflammation including eosinophils. There was also evidence of scarring of the septa between subcutaneous lobules of fat. She was placed on prednisone 30 mg which was tapered to 10 mg over the next month. Her prednisone was tapered again and stopped over the next 4 months, during which time the swelling in her leg gradually went away.

**Discussion:** Eosinophilic fasciitis causes cutaneous changes including pitting edema and irregularity of the skin surface as a fine *peau d'orange*. Several etiologic agents have been described as causing eosinophilic fasciitis, including arthropod bites and borreliosis. There are no known reports of cases of fire ant bite-induced eosinophilic fasciitis.

**C13 A CASE OF ACQUIRED FACTOR VIII INHIBITOR PRESENTING POST-PARTUM**

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**Case Report:** A 37-year-old Vietnamese female patient gravida 3 para 3 without a significant past medical history presented with bruising of the right lower extremity. She had undergone an uneventful C-section two months prior. Laboratory evaluation revealed an elevated partial thromboplastin time (aPTT), which was 101.5 seconds (normal: 23.0-39.1 secs). Mixing studies were consistent with a factor inhibitor presence. The patient's Factor VIII activity level was decreased to less than 0.25%. Factor VIII inhibitor level was significantly elevated at 80 BU (normal: 0-0.5 BU). Based on these findings, the diagnosis of acquired Factor VIII inhibitor (also referred to as acquired hemophilia related to pregnancy) was made. The patient refused further treatment until she presented to the emergency room with hemoglobin of 2.6 gm/dl (12.0-16.0) and worsening bruising approximately one month after diagnosis. Treatment with recombinant Factor VIIa (rFVIIa) 4800 µg IV every 2 hours for a total of four doses and oral steroids were administered. Subsequently cyclophosphamide 100 mg daily was added. The patient clinically improved and her Factor VIII inhibitor became zero. Acquired hemophilia secondary to acquired Factor VIII inhibitors has potentially fatal complications, with mortality rates as high as 22%. The case presented represents an extremely rare condition with a prevalence rate of 0.2 to 1 per million individuals for acquired Factor VIII inhibitors, with only 7%-11% occurring in pregnancy or postpartum states. This case demonstrates a rare and serious bleeding disorder requiring prompt clinical recognition followed by initiation of therapy to reduce mortality.

**C15 COMPLETE RESPONSE OF GLIOBLASTOMA TO CONCURRENT TEMOZOLOMIDE AND RADIATION THERAPY**

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**Background:** To report a complete response to chemoradiation in a patient with glioblastoma multiforme (GBM). GBM is the most common and lethal brain cancer, with a median survival of 12 months after surgery and radiation. Concurrent temozolomide (TMZ) and external beam radiation therapy plus six cycles of TMZ has become the current standard of care for patients with GBM due to better outcome measures like median survival and 6-month progression-free survival. However, a complete radiological response is exceptionally rare, so radiographic responses are no longer outcomes in phase II-III trials.

**Case Report:** A 59-year-old woman presented with a 3-week history of headaches, dizziness, nausea, and speech disturbance. An MRI showed a 2.3 cm ring-enhancing mass in the right temporal lobe. She had a subtotal resection, followed by external beam radiation therapy (60 Gy for 6 weeks) and TMZ (75 mg/m<sup>2</sup> of body surface area x 42 d). She tolerated this regimen well, with mild fatigue and nausea. An MRI of the brain one month after radiation showed no evidence of residual disease on contrast. The vasogenic edema present in T2 and FLAIR sequences had also disappeared.

**Conclusion:** Complete responses in treatment with this regimen have been reported for malignant gliomas and metastatic melanoma but not with GBM, indicating that this is an exceptional event. It also shows that GBM can be chemosensitive, probably due to the expression of methylated O6-methylguanine-DNA methyltransferase (MGMT).

**C16 DONOR-TRANSMITTED MALIGNANT MELANOMA IN A LIVER GRAFT RECIPIENT**

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**Case Report:** Transmission of donor-derived malignancy to the recipient is a rare complication of cadaveric solid organ transplant. This report describes the presentation and outcome of a male recipient who developed malignant melanoma after undergoing a liver transplant from a 53-year-old female donor. Six months after his transplant, the patient developed persistent mild abdominal pain, malaise, and peripheral edema. Work-up included a CT scan, which demonstrated complete replacement of liver parenchyma with a heterogeneous mass. Liver biopsy was positive for metastatic malignant melanoma. No primary source or extra-hepatic uptake was identified on PET scan. Suspicion of donor-derived malignant melanoma was confirmed by utilizing fluorescent in situ hybridization, which demonstrated an overwhelming XX chromosome pattern within the tumor cells. The recipients of the donor kidneys from the same donor showed no radiological evidence of donor-derived malignancy at the time of diagnosis. One recipient underwent transplant nephrectomy that demonstrated no microscopic evidence of neoplastic disease. The other declined nephrectomy and currently remains on immunosuppression without evidence of neoplastic disease. Modern advances in transplant medicine have raised the upper age limits of donors entering into the donor pool. This expansion increases the number of available organs, but it also raises the likelihood of complications due to the age of the donated organs. This is a potentially important issue that needs to be addressed as older individuals enter into the donor pool.

**C17 POSTERIOR REVERSIBLE ENCEPHALOPATHY SYNDROME ASSOCIATED WITH TEMOZOLOMIDE**

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**Background:** Temozolomide (TMZ) is now the drug of choice to treat malignant gliomas. Headaches and seizures are known and adverse effects have been noted in clinical trials, although drug causation is unproven for many of these symptoms. We report an adverse drug reaction to TMZ.

**Case Report:** A 19-year-old man with primary diffuse meningeal gliomatosis began treatment with adjuvant TMZ (150 mg/m<sup>2</sup> days 1-5, every 28 days), preceded by craniospinal irradiation with concurrent TMZ (75 mg/m<sup>2</sup> orally). On day 3 of treatment, he developed headaches, confusion, and seizures. On admission, his blood pressure was 141/105mmHg and was delirious. A brain MRI on admission was compared with a baseline MRI taken two days before TMZ treatment, showing bilateral subcortical and cortical lesions in parietooccipital and posterior frontal lobes with increased apparent diffusion coefficients. We stopped TMZ and started levetiracetam 250 mg twice daily. Three days after admission the patient was clinically better and was discharged. We followed him one, three, and eight weeks after discharge. His mental status improved but never returned to baseline. We restarted TMZ for cycle two at 100 mg/m<sup>2</sup>. A follow-up MRI six weeks after admission showed complete disappearance of the hyperintense lesions. The patient continued treatment with TMZ but had disease progression and died seven months after admission. The posterior reversible encephalopathy syndrome (PRES) is the acute and variable presentation of headaches, delirium, seizures, and visual deficits associated with bilateral cortical and subcortical vasogenic edema predominantly in the posterior areas of the brain. The most common causes of PRES are hypertensive encephalopathy, eclampsia, and immunosuppressive drugs in transplant patients. PRES has been described in adult and pediatric cancer patients treated with chemotherapy. Complete resolution of symptoms is the rule after the causative drug is stopped, but there are exceptions.

**Conclusion:** Temozolomide was directly associated with PRES in this patient because of the following: the close temporal relation between onset of treatment with temozolomide and symptoms without any other modification to the patient's drug list; the radiographic changes by MRI taken before chemotherapy and during admission days later; the resolution of the syndrome after withholding TMZ; and the growing association between cytotoxic drugs and PRES.