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JFK MEDICAL CENTER

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New Jersey Neuroscience Institute at JFK Medical Center

New Jersey Neuroscience Institute (NJNI) at JFK Medical Center is a comprehensive facility designed exclusively for the diagnosis, treatment, and research of complex neurological and neurosurgical disorders in adults and children. Services offered at the Institute include programs in minimally invasive and reconstructive spine surgery, peripheral nerve surgery, brain tumors, dizziness and balance disorders, epilepsy, sleep, memory problems/dementia, cerebral palsy, stroke, spasticity, movement disorders, and neuromuscular disorders. As a department of Seton Hall University's (SHU) School of Graduate Medical Education, NJNI serves as the clinical setting for residency training in neurology and fellowship training in clinical neurophysiology and sleep medicine. For more information on the New Jersey Neuroscience Institute, call 732-321-7950 or visit the facility online at www.njneuro.org.

AIM and SCOPE

The *Journal of the New Jersey Neuroscience Institute* (JNJNI) focuses on topics of interest to clinical scientists covering all subspecialty disciplines of neuroscience as practiced in the Institute and makes clinical information accessible to all practitioners. The fundamental goal is to promote good health throughout the community by educating practitioners and investigating the causes and cures of neurological and neurosurgical ailments.

JNJNI publishes the following types of articles: editorials, reviews, original research articles, historical articles, controversies, case reports, what's new in neuroscience, images in neuroscience, letters to the editors, and news and announcements.

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Editors' Corner

JNJNI, first published in 2007, encourages submissions of clinically relevant articles to enhance the quality of patient care and excellence of the journal. We begin this issue by trying to rejuvenate you, the readers and the contributors. This is your journal and its success belongs to all who are dedicated to raising the standard of the journal and, indirectly, patient care. We hope to attract quality articles from clinicians and researchers both within and outside the Institute in future issues. This is a call for action.

In this winter issue we are publishing five articles in addition to our ongoing section “What’s New in Neuroscience?”

The first article is a timely review of low back pain management. This is probably the commonest condition in neuroscience practice, and it requires meticulous attention. Karnaugh and Vaid write an excellent review of management issues, strictly adhering to practical points and emphasizing the clinical approach. Their fundamental point is that management must depend on astute clinical judgment and assessment—not on laboratory investigations (e.g., MRI, EMG/NCV, etc.)—in order to provide the best quality treatment, rather than subjecting patients to a battery of tests and numerous unnecessary drugs, some of which are very toxic and addictive.

The second article is a case report with a logical approach to a complicated and unusual case of chiari malformation with brainstem compression and Creutzfeldt-Jacob disease by Neiman and coinvestigators. The authors have shown that through a structured approach, rather than haphazard investigations, one can make a diagnosis even in a complex and rare case.

The third article by Farheen describes an interesting case of monomelic amyotrophy which often poses a clinical dilemma in terms of differential diagnosis between the relatively benign and more serious conditions like amyotrophic lateral sclerosis.

The fourth article deals with an uncommon condition, namely rapid eye movement (REM) sleep predominant obstructive sleep apnea (REM-pOSA), which may have serious long-term adverse consequences (as in OSAS) particularly on the cardiovascular system. Many sleep specialists, including neurologists, are not always aware of this.

The fifth article appearing under the “Historical Section” gives a fascinating overview of legal insanity as a legal defense, starting from ancient to modern jurisprudence, citing many historically significant examples.

The final section “What’s New in Neuroscience?” deals with two articles. Dijk and Tijseen gave a nice overview of therapeutic management of patients with myoclonus based on clinical, etiological and anatomical classification of myoclonus. The second article by Dubois and collaborators brought to our attention the revised new definition of Alzheimer’s disease clinical spectrum based on recent advances in the use of reliable biomarkers instead of the time-honored McKhann criteria. This will stimulate further research for potential drug discovery to intercede in the pathogenic cascade of the disease.

We hope these articles will stimulate readers and contributors to look for a copy of *JNJNI*’s last issue of 2010 and encourage new submissions for the spring 2011 issue.

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Management of Low Back Pain

Ronald D. Karanagh, MD; Sandeep Vaid, MD

Abstract

Low back pain (LBP) is a common complaint that prompts many patients to visit their primary care provider and causes a significant socioeconomic burden on the U.S. healthcare system. The management of LBP requires the clinician to be aggressive with the diagnosis, yet conservative with the medical management. A comprehensive patient evaluation is required to identify the pain generator and help guide clinicians toward an appropriate treatment plan. Furthermore, imaging and diagnostic studies should be performed as an extension of the history and physical examination to help identify an underlying pathology and to guide treatment. Most importantly, the clinician should be aware of red flags which may indicate the presence of a more serious pathology requiring immediate intervention. Implementation of a multidisciplinary conservative care approach consisting of reassurance, patient education, recognizing underlying psychosocial problems, NSAIDs, physical therapy, and injection procedures can prevent acute LBP from progressing into chronic LBP.

Introduction

Spine Care is the fastest growing sector of outpatient practice for neurologists, physiatrists and primary care providers. An estimated 25% of adults in the U.S. report at least one episode of low back pain (LBP) in the previous 3 months. LBP is reported to be the fifth most common reason that patients visit their primary care provider in the U.S.¹

The management of LBP requires a comprehensive

approach with the goal of alleviating pain, restoring function, and improving overall quality of life. The clinician must formulate an accurate diagnosis in order to implement a targeted treatment plan. To achieve this goal, the clinician must be able to obtain a detailed history and perform a comprehensive physical examination. The results from the evaluation will then determine the appropriate treatment plan and whether further diagnostic testing is warranted.²

History

LBP is common and generally a benign, self limiting disease. During the initial presentation, the neurologist should rule out any life threatening causes for LBP and be aware of red flags (see Table 1). For example, cauda equina syndrome (which may present as saddle sensory disturbances, bladder and bowel dysfunction, and variable lower extremity motor and sensory loss), malignancy (night pain that awakens the patient from sleep and unexplained weight loss) or spinal infections including discitis, osteomyelitis, and epidural abscess (with associated febrile illness) all require emergent workup and intervention.³ The clinician should make note of symptom onset in order to differentiate acute LBP from chronic LBP. The subjective patient history should include the quality of pain, timing, frequency, severity, referral pattern, aggravating and alleviating factors and any other associated symptoms.⁴ The use of a pain assessment chart (such as a body chart) can be helpful (see Figure 1) in localizing the pain while identifying areas of radiation. It is important to note that by the time a patient presents to the neurology clinic for

low back pain, they have most likely tried conservative methods of treatment. These treatments may include over the counter medications for pain, chiropractic manipulation, physical therapy, and epidural steroid injections. Identification of previous effective and non-effective medications or modalities will aid in the differential diagnosis. Detailed questioning of the patient about aggravating and alleviating factors can assist in localizing the probable pain generator. Pain associated with prolonged sitting, forward flexion, pain radiating down the leg, coughing, sneezing, and bowel movement may be discogenic in nature. Pain that is exacerbated with prolonged walking, standing, spinal hyperextension or lateral bending may be secondary to

facet joint dysfunction.⁵ Patients may also complain of pain at the tailbone with prolonged sitting, standing up from sitting position, and bowel movement, which may be due to coccydynia. Coccydynia can be secondary to trauma, fracture, and malignancy to the coccyx bone. The clinician should, however, keep in mind that these pain generators have overlapping referral patterns and the physical examination will further localize the source of pain.

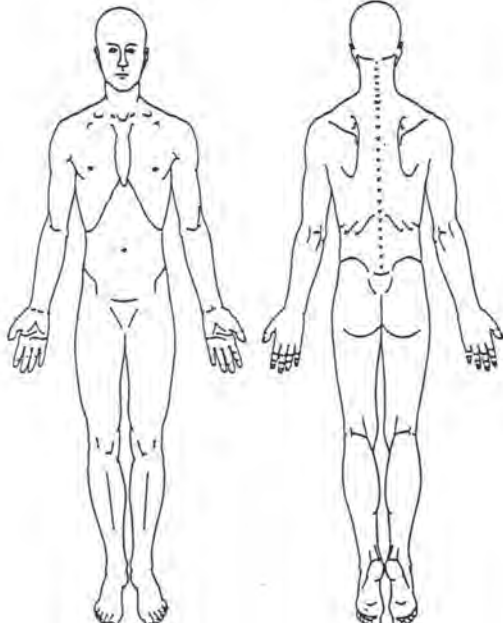
Physical examination

In conjunction with the detailed history, a comprehensive physical examination will assist in narrowing the diagnosis. The examination of the

Figure 1.

Please Complete The Pain Drawing

WHERE IS YOUR PAIN? Please mark on the drawing where you feel pain right now and use the key below the figures.



Pins & Needles - oooo	Burning - xxxx
Stabbing - ////	Deep Ache - zzzz

Figure 1.

Rate Your Pain

0 = no pain 10 = extreme pain

1. Right Now: 0 1 2 3 4 5 6 7 8 9 10
2. At Best: 0 1 2 3 4 5 6 7 8 9 10
3. At Worst: 0 1 2 3 4 5 6 7 8 9 10
4. What makes it better?

5. What makes it worse?

6. If you have low back problems, what percent of your pain is:
(R) Leg: ____% (L) Leg: ____% Back: ____%
(These 3 numbers should add up to 100%)
7. Previous Treatments:

lumbar spine should focus on the evaluation of the musculoskeletal, neurological, and vascular systems. Focus should be placed on several key elements including visual inspection, palpation, range of motion, neurological, and functional assessment.

Pain associated with flexion can be secondary to pathologies of the intervertebral disc, vertebral body, interspinous ligament, or paraspinal musculature. This will help narrow the differential diagnosis to discogenic pathology, compression fracture, interspinous ligament sprain, or paraspinal muscle strain. Typical patient complaints with such diagnoses include pain with sitting, bending or lifting. The pain will improve in supine or prone position or while standing or walking. On examination forward flexion will increase the pain intensity and the patient may complain of radicular symptoms in the leg. Special tests which provoke symptoms such as a straight-leg raise (SLR), slump test, or femoral nerve stretch test will further clarify if the pain generator is discogenic in nature (see Table 2).⁶

LBP associated with back extension is usually associated with posterior element pathology such as pars interarticularis defect, facet arthropathy, foraminal stenosis, or lateral recess stenosis. Patients will complain of pain with standing, walking, running, and extension. The pain is generally relieved when flexing the back, sitting or lying with some flexion, but the pain is typically worse when lying prone. Patients may complain of some leg symptoms, but usually the pain does not radiate below the knee unless significant stenosis is present. On examination, there is increased back pain with extension and lateral bending but no significant pain on forward flexion.⁶ Tenderness to palpation in the region of lower lumbar facet joint is noted and this can be localized close to the sacroiliac (SI) joint. SLR is usually negative; however,

hamstrings are usually tight. Possible generators for back pain associated with hyperextension include spondylosis, scoliosis, spondylolysis, spondylolisthesis, spinal stenosis, and facet joint syndromes.

Confirming the diagnosis

Diagnostic studies should be used as an extension of the history and physical examination and not as a substitute. They should not be ordered indiscriminately due to number of false positive results noted in

Table 1.

Life Threatening and Urgent Conditions
MEDICAL
Infection: osteomyelitis, epidural abscess, discitis
Hematologic: primary or metastatic cancer, multiple myeloma, myelodysplasia
Retroperitoneal pathology: pyelonephritis, renal calculus
Benign tumors
Aortic aneurysm
Abdominal pathology: pancreatitis, perforated viscera
MUSCULOSKELETAL
Lumbar or sacral nerve root compression
Disc herniation, cauda equina syndrome, spinal stenosis
Vertebral fracture
Sacroiliac joint sprain
Arthritic conditions: osteoarthritis, rheumatologic conditions
Lumbar muscle strain

Modified from Deyo et al.³

asymptomatic subjects. Diagnostic studies such as plain radiographs of the lumbar spine are not routinely needed in the evaluation of most episodes of LBP. Abnormalities on X-ray do not correlate with degree of patient symptoms, but X-rays are helpful in the initial evaluation to rule out a more serious condition in patients with red flags such as tumors, osteomyelitis, and retroperitoneal pathology including small and large bowel obstruction and bowel perforation.³

Bone Scans

In general, bone scans are not useful in evaluating acute LBP. However, they can be very helpful in confirming the diagnosis of an acute spondylolysis (stress fracture of pars interarticularis) especially with single photon emission computed tomography (SPECT) imaging. Furthermore, they may be useful when the history and examination indicate the possibility of infection or tumor.⁷

Table 2. Provocative maneuvers for the diagnosis of lumbar disk herniation.

MANEUVER	PROCEDURE
Straight-leg raise	With the patient lying supine, the leg is raised with the knee extended; elevation of the leg is stopped when the patient begins to feel pain; the result is positive when the angle is between 30° and 70° and the pain is reproduced down the posterior thigh to below the knee.
Cross straight-leg raise	Same as above, with pain elicited on raising the contralateral leg.
Slump test	The patient is seated with legs together and knees against the examination table; the patient slumps forward as far as possible and the examiner applies firm pressure to bow his or her back while keeping the sacrum vertical; the patient is then asked to flex the head, and pressure is added to neck flexion; the examiner then extends the knee and adds dorsiflexion to the ankle; the test result is positive when there is reproduction of symptoms.
Femoral nerve stretch test	With the patient prone, the examiner places his palm at the popliteal fossa as the knee is dorsiflexed; the test result is positive when there is reproduction of symptoms in the anterior thigh or back or both; this test is used to make a diagnosis of high lumbar disk herniations.

Modified from Solomon J et al. *Physical examination of the lumbar spine*. In: Malanga GA, Nadler SF, editors: *Musculoskeletal Physical Examination*. Philadelphia: Elsevier Mosby, 2006; p. 210-213. Reproduced with permission from GA Malanga.

Magnetic Resonance Imaging (MRI)

MRI has been shown to have excellent sensitivity in the diagnosis of lumbar disc herniation. This modality should be reserved for patients who present with persistent pain for more than 6 to 8 weeks or with neurologic deficits such as progressive weakness, sensory deficit, or a dropped reflex. Other considerations include cauda equina symptoms, high risk patients for cancer, infection, or inflammatory disorders.⁸ It should be noted, however, that a significant number of false positive results are found in the asymptomatic population.⁹ In patients presenting with pain that radiates in a specific dermatomal distribution along with other neurological findings of weakness, sensory changes, or asymmetrical reflex secondary to a radiculopathy due to disc herniation, an MRI study of lumbar spine is necessary to help guide treatment such as an injection procedure. In the patient presenting with LBP, the addition of gadolinium is not necessary in the majority of patients.⁹

Computerized tomography (CT) imaging

CT scans are a good modality for imaging osseous structures; however, they are inferior to MRI for detecting disc herniation. Patients who are unable to undergo MRI scanning due to permanent pacemaker or metallic implants are imaged using CT. A CT scan of the spine can detect spinal stenosis, facet arthrosis, herniated nucleus pulposus (HNP), spondylolysis, osteoporosis, and neoplasms.¹⁰ CT scans of the spine in conjunction with myelography are a great modality for patients with spinal stenosis who are contemplating surgery.

Electrodiagnosis

Electrodiagnosis, which includes electromyography (EMG) and nerve conduction studies (NCS), should be considered as an extension of the physical examination. It is helpful among patients with limb pain where the diagnosis remains unclear. The study can be helpful in ruling out other causes of sensory and motor complications such as peripheral neuropathy, mononeuropathy, and motor neuron disease. EMG studies can be used in patients with significant weakness secondary to neuropraxia and pain inhibition from significant axonal injury. The use of EMG is not necessary in patients with isolated low back symptoms. If diagnosis of radiculopathy is unequivocal after a detailed history and physical examination, the addition of EMG and NCS does not improve the treatment outcome and thus is not required.¹¹

General treatment principles

LBP treatment should begin once the diagnosis is made and the red flags have been ruled out. LBP management requires a multifaceted approach with the goal of minimizing pain, normalizing activities of daily livings, and improving overall quality of life. The natural history of LBP is to improve with or without treatment, but certain treatments can hasten the process and are worthwhile. Successful management requires the patient's participation in the treatment plan and understanding the cause of the LBP. The clinician must explain the working diagnosis and review the basic anatomy and biomechanics that led to the LBP. The treatment plan should be reviewed and rationale for diagnostic studies should be identified. It is important to note that the vast majority of patients with LBP can be treated through conservative methods with reassurance. However, patients presenting to the neurologist or physiatrist may have tried conservative

methods without success. These patients may be candidates for more aggressive treatment modalities such as stronger non-steroidal anti-inflammatory drugs (NSAIDs), opioid analgesics, oral corticosteroids, antidepressants, antiepileptic medication, injection therapy, and finally surgical intervention. Again, the physician must have the correct diagnosis in order to implement the correct treatment plan.

Life style modification

For most patients, LBP can be treated with lifestyle modification and education. Patients should be instructed on proper posture and biomechanics while performing household and occupational activities. The American Obesity Association has reported that one-third of Americans classified as obese suffer from musculoskeletal and joint pain. Due to the excess weight the spine can become misaligned and stressed unevenly. Weight reduction has been proven to control pain among this population. Studies have shown that smoking is also a causative factor of LBP. A recent publication by Shiri and associates concluded that current smokers and individuals who smoked in the past have a higher incidence of LBP when compared to individuals who never smoked.¹² The risk of back pain associated with smoking was modest among adults and was noted to be even greater in the adolescent population. Lastly, one must also consider assessing psychosocial factors which will undermine the results of treatment, i.e., by treating depression in conjunction with treating the patient's underlying pain generator.

Bed rest

Some benefits can be gained from bed rest secondary to the reduction in intradiscal pressure while lying in the supine position. But bed rest has many detrimental effects on bone, connective tissue, muscles, and

cardiovascular fitness. For non-radicular LBP, 2 days of bed rest have been shown to be as effective as 7 days.¹³ For radicular symptoms, however, limited bed rest in conjunction with standing and walking (as tolerated) is beneficial and ideal.¹⁴ Patients should be educated on positions that may exacerbate their back pain symptoms. Patients with discogenic pain should be instructed to avoid prolonged sitting, bending and lifting and educated on proper work station ergonomics to minimize back and neck pain due to poor posture.

Exercise

The goal of an exercise program in patients with LBP is to control pain, strengthen muscles and restore proper motion of the spine and trunk. Studies have shown that patients with LBP have a reduction in aerobic fitness that may contribute to exacerbation of pain.¹⁵ Conversely, evidence has shown that athletes with high cardiovascular fitness rarely have LBP.¹⁶ Improving cardiovascular fitness in addition to an active exercise program is a reasonable treatment modality; patients should be encouraged to stay active in order to avoid deconditioning.

Physical therapy

There is conflicting literature on the effects of strengthening exercises in acute LBP, but their effects on patients with chronic LBP have the best outcome. Various exercise modalities are used when strength training such as flexion, extension, and dynamic lumbar stabilization exercise. Debate surrounds the merits of flexion versus extension type exercise. Flexion exercise appears to be more reasonable in patients with posterior element problems;¹⁷ extension exercise has been effective in patients with discogenic LBP.¹⁸ A dynamic lumbar stabilization program has been shown to be most effective for a multitude of low

back problems.¹⁹ The program emphasizes maintaining a neutral spine position coupled with progressive strengthening exercise of the trunk muscles, i.e., back extensors, abdominals, and gluteal muscles. The goal is to develop muscular support of the trunk to diminish stress on the bones, discs, ligaments, etc.¹⁹ This treatment modality requires close supervision, direction and a hands-on approach by the treating physical therapist.

Manipulation/Mobilization

With treating acute LBP, several studies suggest that manipulation during the first 3 weeks decreases painful episodes, but this is controversial. Nevertheless, the benefits are primarily reduction in symptoms in the acute phase with no evidence of long term benefits. Only patients with LBP appear to respond; those with radicular pain do not show improvement in their pain.

Medications

Medications from many different classes have been used to treat LBP and each has unique risks and benefits in the treatment. When prescribing these medications, the clinician should have knowledge of the indication and contraindication.

Acetaminophen

Acetaminophen is an inexpensive over the counter medication and generally a safe analgesic. It is effective for mild to moderate pain but has no effect on inflammation or muscle spasm. This medication is generally not considered a first-line medication for LBP and used if the patient has contraindication to other medications.

Nonsteroidal anti-inflammatory drugs (NSAIDs)

NSAIDs are reasonable medications for pain relief and have anti-inflammatory effects. NSAIDs are most effective during the first week of exacerbation and the anti-inflammatory doses are significantly different from the analgesic dose. Many times, the doses prescribed are too small and far too short to produce an anti-inflammatory effect.²⁰ There are high risks associated with NSAIDs, particularly in elderly patients and patients with hypertension, diabetes, and gastrointestinal ulcer; therefore prolonged use (greater than 4 to 6 weeks) should be avoided.

Muscle relaxants

Muscle relaxants have the unique effect of inducing muscle relaxation and often sedation; they primarily affect the central nervous system (CNS) and have associated effects on the neuromuscular system. They are often prescribed during episodes of acute LBP and have been shown to be beneficial when used with NSAIDs. Muscle relaxants should be used at night to take advantage of their sedating effect and to minimize daytime sedation.

Opioid analgesics

Use of opioid analgesics in acute LBP should be limited and considered only in patients with pain that is unresponsive to NSAIDs and muscle relaxants. When prescribed for chronic LBP, they should be written on an appropriate dosing schedule and not on a PRN basis.²¹ If opioid medications are used, the risks and benefits should be considered due to potential addiction or misuse in patients with abuse history.

Oral corticosteroids

Theoretically these agents are useful in patients with radiculopathy due to disc herniation. But there are no controlled studies to support their use, and only anecdotal clinical success is reported.

Antidepressant medication

Antidepressant medications have been shown to be helpful in the treatment of chronic LBP. These medications can typically take up to 4 weeks to be effective and studies suggest that the effects are not dependent on changes in depression scores.²² Antidepressants are usually not indicated in the treatment of acute LBP.

Conservative modalities

Transcutaneous electrical stimulation (TENS)

TENS is a modality used to treat a variety of pain conditions and generally used in the treatment of chronic pain conditions. These modalities are generally not indicated in the treatment of acute LBP and the success rates vary greatly. The mechanism of pain reduction is secondary reduction in nerve conduction, muscle contractility and its counter-irritant effects.²³ Use of electrical stimulation should be limited to the initial phase of treatment to facilitate an active exercise program.

Ultrasound

Ultrasound is a deep heating modality and most effective in heating deeper musculoskeletal tissue. It improves connective tissue distensibility that facilitates stretching of contracted tissue. This modality is often misused and abused due to its use in acute injuries and especially in acute radiculopathy. Ultrasound as a therapeutic modality should be used to facilitate soft tissue mobilization and to improve range of motion.

Superficial heat

Heat modality is helpful in decreasing stiffness in smaller more superficial joints. Heating effects occur at a level of 1 to 2 cm and can decrease pain and muscle spasm. This modality should be used as an adjunct to facilitate an active exercise program.

Cryotherapy

Cryotherapy is a more effective tissue penetration modality which is able to decrease muscle temperature by as much as 3 to 7°C. This causes vasoconstriction which in turn results in a reduction of metabolism, edema, and local inflammation. There is a decrease in nerve conduction velocity and reduction in muscle spasm. Cryotherapy treatment should be employed 15 to 20 minutes, 3 to 4 times a day during the acute injury.²⁴

Therapeutic injections

Therapeutic injections are helpful in confirming a diagnosis after a careful history and physical examination. This interventional modality should not be used in isolation and the number should be limited to that which allows for an active exercise program. Therapeutic injections can range from in office trigger point injections to minimally invasive epidural steroid and facet joint injections performed under fluoroscopic guidance.

Trigger point injection

Myofascial trigger points are felt to be hyperirritable foci in muscle associated with taut muscle bands. They are diagnosed by palpation and production of local pain that is referred away from the site of the tender muscle. The injection of these foci should be reserved for patients who have not responded to other treatment modality after 4 to 6 weeks. There is no

evidence to support the use of corticosteroids in these injection techniques.²⁵ More than 1 injection may be required but greater than 3 in the same trigger point is not usually necessary.

Epidural Corticosteroid Injection (ESI)

The rationale for the use of epidural injections has improved with the evidence of inflammatory mediators in patients with radiculopathy. The physician must correlate the patient's history, physical examination findings, and imaging studies to determine that the appropriate procedure is indicated for further diagnostic and therapeutic value. The goal of ESI is to reduce the pain and inflammation so the patient can progress to an active exercise program.²⁶ There is release of inflammatory mediators in patients with disc herniation and acute radiculopathy which leads to pain and immobility. In our clinical practice, all patients who present with radiculopathy (except cauda equina syndrome) receive a trial of ESI before any surgical

referral is made. Scientific evidence regarding efficacy is mixed, but overall the short term results have been very positive in patients with acute radiculopathy from disc herniation. In a prospective case series which examined the outcomes of patients with lumbar HNP and radiculopathy who received transforaminal epidural steroid injections (TF ESI), Lutz et al²⁷ found that 75.4% of the patients had a successful long-term outcome, as determined by their post-injection pain scores and ability to return to previous levels of functional activities. In summary, TF ESI is the treatment of choice for unilateral radicular pain (see Figure 2).

In our clinical practice, many patients with spinal stenosis have benefited from caudal ESI. Fluoroscopic guidance is imperative for proper placement and administration of the medication. Repeat injection should be based on pre-treatment goals and the therapeutic response. It is not necessary for most patients to undergo a set number or series of injections.

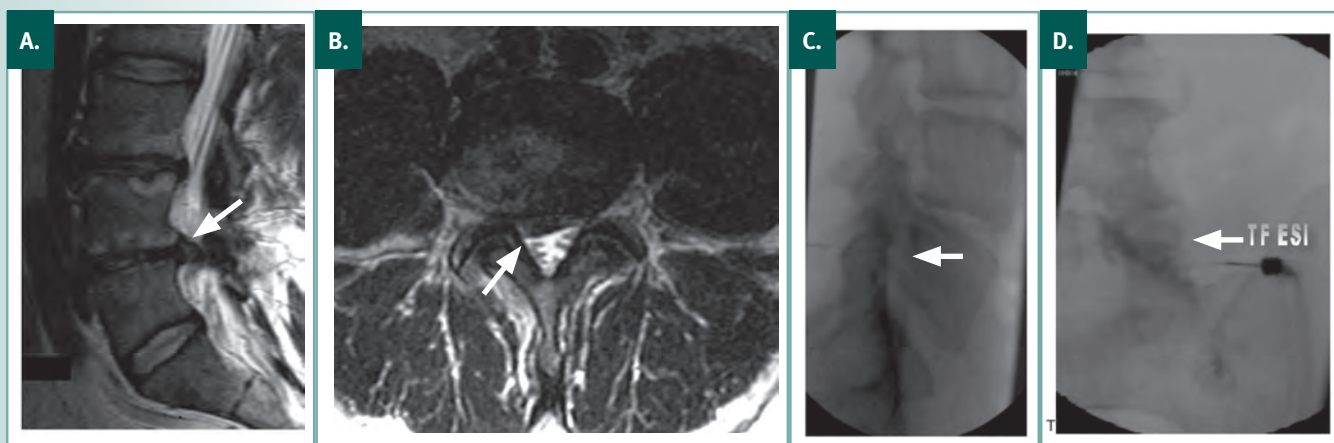


Figure 2. A 44-year old male with a 3 month history of severe LBP with radiation to right leg; physical examination findings and imaging studies (lumbar MRI above) correlated with diagnosis of right L5 radiculopathy secondary to L4-5 disc extrusion. He underwent a right L5 Transforaminal Epidural Steroid Injection (TF ESI) and at follow up remains asymptomatic. A: Sagittal view shows evidence of a large L4-5 disc extrusion. B: Axial view confirms a large right L4-5 paracentral disc extrusion (measuring approximately 8mm in size) which narrows the right lateral recess and causes a mass-effect on the traversing L5 nerve root in the spinal canal. C: Lateral view of a fluoroscopically guided, contrast enhanced right L5 TF ESI with needle tip placement into the superolateral neuroforamen. D: AP view with needle tip placement under the 6 o'clock position of the L5 pedicle with contrast dye flow outlining the exiting L5 nerve and into the proximal epidural space.

If no improvement in pain is noted after two injections, then a third injection is not indicated. Patients should be followed up after the injection to monitor pain level and reassess neurological status.

Facet injections

Facet joints are a potential source of LBP generation and the facet injection is used in the diagnosis of facet mediated pain after correlating the history and physical examination findings with imaging. The facet injection should be reserved for patients with examination findings consistent with facet pain who have not responded to treatment in the first 4 to 6 weeks.²⁵ Even though facet joints can be a source of pain, the therapeutic benefits of the injections are controversial.

Conclusion

Management of LBP requires the clinician to be aggressive in the diagnosis and conservative in the management. The neurologist must be knowledgeable in the anatomy and the biomechanics of the spine to assist in the diagnosis of LBP. Obtaining a comprehensive history and performing a thorough physical examination will lead to differential diagnosis which can be further confirmed by imaging studies, if needed. The implementation of the treatment plan requires that the proper diagnosis for LBP has been made. Understand the Natural History and intervene when you can change it with the least invasive options first. The clinician should keep in mind that most patients with LBP will improve over time and that they should be active and not become bed bound. Treatment comprises of reassurance, education with life style modifications, and addressing underlying psychosocial issues. Literature has shown that also starting a short course of NSAIDs and an active exercise program

at the initial onset of LBP will allow the patient to return to normal activity and prevent the back pain from becoming chronic. LBP in some patients will become chronic in nature and these patients will benefit from a dynamic lumbar stabilization program and core musculature strengthening to help control and prevent further back pain and deconditioning. For patients with back pain secondary to mechanical and degenerative pathology, minimally invasive approaches with injections will help control and alleviate LBP with or without radiculopathy. Currently in our clinical practice, all patients without cauda equina receive a trial of conservative treatment and if they do not respond then a surgical referral is made. We will continue to treat the patient, and not the MRI findings. Our goal is to assess and treat the patient with acute LBP appropriately and hence prevent creating the patient with chronic LBP.

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An Unusual Presentation of Creutzfeldt Jacob Disease and An Example of How Hickam’s Dictum and Ockham’s Razor Can Both Be Right

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Introduction

Patients can have more than one neurological problem, and sorting out acute from chronic disease can be challenging. We report a middle-aged patient who presented with ataxia, right hemiparesis, and abnormal nystagmus. Magnetic resonance imaging (MRI) showed a Chiari and an arachnoid cyst with brainstem compression that appeared to explain his abnormal examination. Shortly after admission he was noted to have intermittent abnormal behaviors and confusion. History from family revealed significant acute and chronic psychiatric problems that appeared to explain his abnormal mental status; this delayed the diagnosis of intermittent complex partial seizures. All of these problems resulted in a delay of the final diagnosis of Creutzfeldt Jacob disease, which in retrospect explained the entire new physical examination, seizures and mental status changes.

Case

A 48-year-old man with mild mental retardation and history of controlled hypertension was escorted to the Emergency Department by the local police after being stopped for driving erratically and being slow to respond to questioning. MRI of the brain showed tonsillar herniation with large left posterior fossa arachnoid cyst. The patient was transferred to our facility for a neurosurgical evaluation concerning possible herniation.

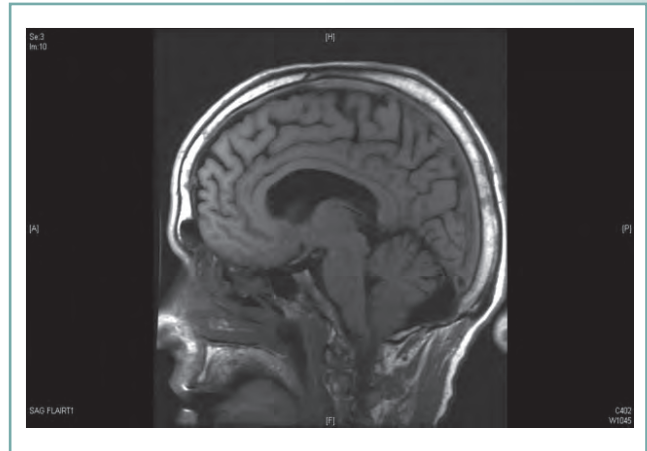


Figure 1. Sagittal FLAIR: Arachnoid cyst and Chiari type 1 malformation.

The family stated that he always had “learning problems,” but for the past month he had become progressively more forgetful, with increasing difficulty in understanding simple commands and a gait disturbance. Behavioral changes were also noted at his place of work. On examination the patient was tearful and confused. Mental status examination revealed disorientation to place, perseveration, and difficulty understanding simple commands. Cranial nerve examination revealed a primary position right beating nystagmus. He had a mild right-sided hemiparesis with intact sensation to all modalities. His gait was ataxic and there was a left-sided dysmetria.

MRI revealed a Type I Arnold-Chiari malformation and a large left lateral posterior fossa arachnoid cyst with compression of the brainstem and tonsillar herniation (Fig 1.) An external ventricular drain (EVD) was

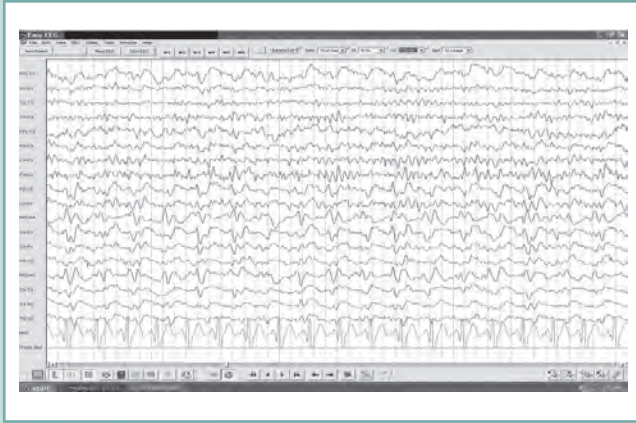


Figure 2. Initial EEG with right hemispheric PLEDs 1.5-2 Hz discharges maximally seen right frontal.

placed into the posterior fossa arachnoid cyst for decompression and to safely obtain a cerebrospinal fluid (CSF) sample without worsening the herniation (Fig 2). No clinical improvement was noticed after decompression. CSF protein, glucose and white blood cell (WBC) count were unremarkable (RBC 707, WBC 4, protein 19.8, glucose 91.7), and CSF cultures for viruses, bacteria, and fungi were negative.

The patient became progressively more agitated and confused. His change in mental status prompted an electroencephalography (EEG), and CSF from EVD was sent. EEG showed 1 to 2 Hz periodic lateralized epileptiform discharges (PLEDs) predominantly in the right hemisphere seen maximally over the right frontal head region (Fig. 3). Electrographic seizures



Figure 3. Burst suppression pattern with generalized periodic epileptiform discharges seen every 1-2 seconds.

were also seen with right frontal onset and spread to the contralateral hemisphere. The patient developed nonconvulsive status. The seizures were difficult to control and were treated aggressively with various anticonvulsants including levetiracetam, phenytoin, valproate, and lacosamide. The patient's seizures became more frequent, requiring intubation, and propofol drip and lorazepam drip were also used to stop nonconvulsive seizure activity. The EEG pattern evolved over weeks to a burst suppression pattern with continuous 1 Hz generalized periodic epileptiform discharges seen every 1 to 2 seconds (Fig 4).

A subsequent MRI showed diffusion weighted imaging abnormality within the basal ganglia (caudate and putamen) and cortical ribbon diffusion restriction

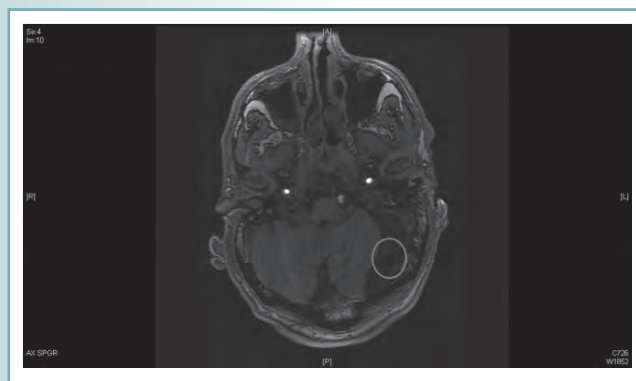


Figure 4. External ventricular drain placed in arachnoid cyst for CSF removal and brain stem decompression.

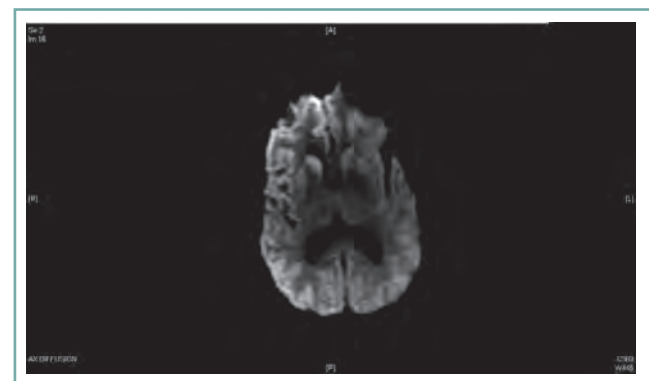


Figure 5. Final DWI: with cortical ribbon sign and more prominent basal ganglia diffusion abnormality.

seen most prominently in the right frontal head region and in the right intrahemispheric region as often classically seen in CJD (Fig 5). He failed weaning trials for extubation and the family withdrew care, per the patient's wishes in his living will. The patient was discharged to hospice where he subsequently died days later and had an autopsy.

The CSF sample sent for protein 14-3-3 was positive. ELISA immunoassay for tau protein was also positive in an amount of 10090pg/ml in CSF (decision point 1200pg/ml).¹ Brain tissue was sent to the National Prion Disease Pathology Surveillance Center in Cleveland, Ohio. Western Blot analysis on frozen sections revealed the presence of abnormal protease resistant prion protein (PrPSc) often identified as PrP 27-30. Immunostaining with 3F4, the monoclonal antibody to the prion protein, revealed granular deposits as seen in prion diseases. The cause of death was sporadic Creutzfeldt-Jakob disease (sCJD) MM1 according to the classification proposed by Parchi et al.²

Discussion

When considering a patient's possible diagnosis there are often discussions about whether or not all of the patient's problems can be explained by a single diagnosis or if in fact there may be several. In medicine it has become axiomatic that a single diagnosis, if possible, is usually the correct one. This philosophy has long been attributed to a 14th century theologian, Father William of Ockham, who said, "plurality should not be posited without necessity."³ (Most medical students and residents of this century have been taught the importance of trying to come to a single diagnosis of their patient's problem and have heard this philosophy attributed to Ockham. It is called Ockham's razor:

diagnoses are "shaved off" the list, leaving the only true diagnosis.

This approach, however, did not translate in our patient. The chiari and arachnoid cyst appeared to explain the physical examination findings, and the psychiatric history similarly seemed compatible with his change in mental status; even the intermittent complex partial seizures seemed to be unrelated. Thus we felt that Ockham's razor had given way to Hickam's dictum. John Hickam, a physician at Duke and then Indiana University, stated (paraphrased) "a man can have as many diseases as he darn well pleases."⁴ Over the next several days, as the patient continued to rapidly deteriorate and an EEG confirmed PLEDS and nonconvulsive status, it became clear that both dictums were true. CJD was a single disease that best explained all features of the patient's subacute course as expected by Ockham's razor. Nonetheless, the patient did have multiple other problems confirming Hickam's dictum.

Our case shows that these ideas are not necessarily mutually exclusive. Although it has become axiomatic that we try to apply Ockham's razor to the differential diagnosis of our patients, we must be careful as some have both "fleas and lice."

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Hirayama disease/Monomelic Amyotrophy: A case report

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Introduction

Monomelic amyotrophy (MMA) is a rare disorder presenting with atrophy and weakness restricted to one limb. It is restricted to lower motor neurons. The benign nature of MMA helps distinguish it from other lower motor neuron disorders like amyotrophic lateral sclerosis (ALS).

Case description

A 26-year-old right-handed Indian man initially noted mild left hand weakness while playing volleyball. Over the next 7 years he developed slowly progressive weakness and atrophy of the entire left upper extremity. He denied pain, numbness, diplopia, dysphagia, ptosis, muscle cramps, fasciculations, headache or neck pain. There was no history of febrile illness, poliomyelitis, and exposure to toxins or heavy metals. He had no significant past medical or surgical history, and the family history was unremarkable. On examination he was alert and oriented to time, place and person. His cranial nerve and sensory examination was normal. Motor examination revealed atrophy of entire left upper extremity and weakness (power of 4/5) of left deltoid, biceps, triceps, wrist flexio and extensor, and hand muscles. The rest of the motor examination was normal. Coordination and gait testing was unremarkable. Serum electrolytes, urea, creatinine, liver function test, thyroid panel, erythrocyte sedimentation rate (ESR), and creatine phosphokinase were normal. Human immunodeficiency virus (HIV) test was negative. Magnetic resonance imaging (MRI) of the cervical

spine showed vertically oriented signal alteration in left mid anterior spinal cord from C3-C7, suggestive of cord atrophy on the left and small disc herniation at C5-C6 on the right. Electrodiagnostic testing revealed normal nerve conduction studies and positive sharp waves and fibrillation potentials on needle electromyography (EMG) testing in almost all of the muscles in the left upper extremity. Fasciculation potentials and motor unit action potentials of increased amplitude and duration with reduced recruitment were also noted in most of the left upper limb muscles tested. The needle EMG of the other limbs was normal. Genetic testing for spinal muscular atrophy (SMA) was negative. The patient was diagnosed with Monomelic amyotrophy (MMA).

Discussion

Benign Monomelic Amyotrophy is a rare condition in which neurogenic atrophy is restricted to one limb.¹ An upper limb involvement is referred to as Brachial Monomelic amyotrophy (BMMA) or Hirayama disease,² and lower limb as crural monomelic amyotrophy.³ MMA is a rare disorder predominantly affecting young men in the second and third decades.⁴ Most of the cases reported are Japanese, Indian or Malaysian.¹ MMA has insidious onset. Sporadic occurrence of wasting and weakness confined to one limb, initial slow progression for 2 to 4 years followed by a stationary course, and absence of spreading to other limbs are characteristic features. There is lack of involvement of the cranial nerves, cerebrum, brain stem or sensory

nervous system.⁵ In upper limb involvement wasting and atrophy of the muscles of medial aspect of the forearm and the small muscles of the hand are noted.⁵ Sparing of brachioradialis muscle is noteworthy in patients.^{5,6} In the lower extremities atrophy is mainly restricted to quadriceps; however, diffuse wasting of entire lower extremity has also been reported.⁵ Irregular, jerky coarse tremors not peculiar to this disease have been reported and designated minipolymyoclonus.^{5,7} Another characteristic feature is aggravation of symptoms on exposure to cold—cold paresis—and it is possibly due to an autonomic disturbance or sensitivity of atrophic muscles to cold.⁵ Interestingly, even after a duration of illness ranging from 5 to 15 years there is no clinical evidence of it spreading to other limbs.⁵ EMG studies are characterized by potentials of increased amplitude and polyphasic potentials consistent with the underlying pathophysiologic process of denervation and reinnervation.¹ Imaging of cervical and lumbar cord has shown variable results: focal unilateral atrophy or anterior horn cells.¹ Computed tomography (CT) and MRI of skeletal muscles can offer useful additional information on muscle involvement.^{1,8,9,10} The differential diagnosis of MMA includes distal spinal muscular atrophy, amyotrophic lateral sclerosis, chronic focal myositis, late progression of poliomyelitis, post polio syndrome, and multiple motor neuropathies.¹ In conclusion, we should consider the diagnosis of MMA in cases of slowly progressive unilateral amyotrophy restricted to one limb followed by stabilization and with neurogenic changes in the EMG.¹

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Cardiovascular Comorbidities in Rapid Eye Movement Sleep Predominant Obstructive Sleep Apnea (REM-pOSA): A Pilot Study

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Abstract

We compared cardiovascular comorbidities in patients with REM-pOSA and those with OSA in general. We retrospectively reviewed 200 consecutive polysomnograms to identify REM-pOSA and selected 15 patients. Five patients with REM-pOSA had a history of hypertension, 2 had diabetes, 4 had hyperlipidemia, and 3 complained of palpitations. These numbers were 5, 2, 5 and 2 patients, respectively, in patients with OSA in general. Thus, there was no significant difference between patients with REM-pOSA and those with OSA in general with regard to hypertension, diabetes, or hyperlipidemia. Nevertheless, palpitations were 1.5 times more prevalent in the REM-pOSA group. The findings suggest that REM-pOSA may represent an early and perhaps milder stage of OSA, and vigorous control of comorbid risk factors may be beneficial in this patient population.

Introduction

The natural history of REM-pOSA is ill defined. Our aim was to study cardiovascular comorbidities in patients with REM-pOSA to determine whether patients with this condition had a significantly different clinical profile from those with OSA in general. We also looked at body mass index (BMI) and demographic features, drawing from patients referred to a single laboratory.

Methods

We retrospectively reviewed 200 adult polysomnograms performed at JFK Medical Center to identify REM-pOSA. A study was included if REM comprised $\geq 10\%$ of total sleep time, the overall apnea-hypopnea index (AHI) was ≥ 5 and the ratio of the AHI in REM (AHIREM) and in NREM (AHI NREM) > 2 . Sleepiness, sleep maintenance and cardiovascular comorbidity information was obtained through patient interviews. Cardiovascular comorbidity (including the presence of hypertension, diabetes and hypercholesterolemia) and demographic data were obtained by contacting patients via telephone or during sleep clinic visits.

Results

We included 15 patients in the case group, with a mean age of 42.1 (SD +/- 8.7), a mean BMI of 32.6 (SD +/- 7.9) and a 0.33 female/male ratio. Mean AHI was 15.9/h (SD 8.0); mean AHI in REM sleep 45.5/h (SD 18.1) and mean REM-AHI/NREM-AHI 3.1 (SD 1.1). This was compared to a comparison group of 15 patients with the following characteristics: mean age of 56.6 (SD +/- 11.9), a mean BMI of 28.3 (SD +/- 3.4), and a 0.3 female/male ratio. Mean AHI was 37.4/h (SD 20.7), mean AHI in REM sleep 48.2/h (SD 20.2), and mean REM-AHI/NREM-AHI 1.4. Thirty-three percent of patients had a history of hypertension, and 20% complained of palpitations.

Discussion

Our study showed that patients with REM-pOSA tended to be younger than those with OSA in general; however, the mean BMI was noted to be higher. This appears to suggest that REM-pOSA may represent an earlier stage of the disease. In addition, the prevalence of hypertension, diabetes, and hyperlipidemia were similar in patients with REM-pOSA and those with OSA in general. Hence, early treatment may help prevent adverse cardiovascular outcomes.

Nevertheless, the number of patients in this study is too small to draw any statistically significant conclusions, and further long-term prospective studies, using more objective measures for cardiovascular risk factors, such as hypertension, seem to be warranted to validate these findings.

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Table 1. Demographic and Clinical Data

	Cases (n=15)	Comparisons (n=15)
Demographic Factors		
Age years (SD)	42.1 (8.7)	56.6 (11.9)
Female n (%)	33.3	33.3
BMI	32.6 (7.9)	27.8 (3.4)
Comorbidities (%)		
AHI	15.9 (8.0)	37.4 (20.7)
REM/NREM AHI	3.1 (1.1)	1.4 (0.3)
Risk for OSA	60	33.3
Hypertension	27	26
Diabetes	15	13
Hypercholesterolemia	33.3	33.3
Palpitations	20	13

As shown in Table 1, other comorbidities were also comparable in the two groups. These included diabetes (present in 15% of the cases and 13% of the comparisons) and hypercholesterolemia (present in 33% of both groups). However, patients with REM-pOSA tended to be younger (mean age 42.1 years) compared to those with OSA in general (mean age 56.6 years). The mean BMI was noted to be higher in patients with REM-pOSA (mean value 32.6) compared to the controls (mean BMI 27.8/hr).

A Historical Overview of Legal Insanity

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Abstract

After reviewing the history and evaluation of the concept of “Legal Insanity,” this article presents 15 well-known individuals, from the 17th century to the present, illustrative of the concept. We conclude with a discussion of the clinical, evaluative, and ethical role of the neurosciences in bridging the formidable gap between clinical sciences and the law.

Keywords: Insanity Defense; NGI/NGRI; legal proofs; scientific proof; criminal/clinical responsibility

“In Anglo-American law, a criminal defendant must be sane to be found guilty of the offense(s) with which he or she is charged. If that individual is not sane, he or she must be adjudicated not guilty by reason of insanity.”¹

Although the legal principle enunciated above applies to a recent (2009) Superior Court decision in New Jersey, its underlying principle is very old in the history of the law. Dating back to Biblical times and first formally articulated in the “Wilde Beeste Test” in 18th century England, the concept of a legal “Insanity test” for criminal responsibility has evolved a great deal over the years. The concept presently consists of 5 variants in the United States and other jurisdictions based on English common law.²

To explore the concept of “Legal Insanity” historically and to discuss its present applications to criminal law, this article will (1) review the historical evolution of

the concept of “Legal Insanity,” leading to its current variants and applications; (2) present, summarize, and discuss the illustrative stories of 15 selected well-known individuals for whom “Legal Insanity” was or may have been an issue in their legal defense, whether they were ultimately adjudicated “legally insane”^{*} or not; and (3) discuss issues and dilemmas inherent to the concept of “Legal Insanity,” as well as alternative approaches to the concept, as used in some jurisdictions.

Background and History of the NGI/NGRI* Defense

Public interest in the legal insanity defense is based on curiosity, often fueled by equal proportions of ignorance and anger, as well as on a firm belief that the justice system is flawed. A common public view is that many criminal defendants either attempt to use the insanity defense or are successful in that attempt. This is not true: The insanity defense is rarely attempted and rarely successful. The NGRI defendant rarely walks happily and freely out of the court. Rather, that exculpated individual is generally committed to a secure institution—often a state hospital—for periods of preventive institutionalization that can be lifelong.

In examining the history of “Legal Insanity” and the “Insanity Defense,” we can only guess how ancient civilizations managed that thorny issue. The ancient Greeks showed some compassion toward people who were violent toward others without reason. The Roman successors to the Greeks regarded mental illness as a sign of religious superiority in some circumstances.³

^{*}In this article, the acronyms “NGI” and “NGRI” (“not guilty by reason of insanity”) will be used to describe this legal determination.

Legal treatment of the “criminally insane” did not advance until medieval England. In 1265, 50 years after the signing of the Magna Carta, England’s Lord Bracton wrote that “...an insane person is one who does not know what he is doing, is lacking in mind and reason and is not far removed from the brutes.”³

The connection made with a type of “brute” as a basis for “Legal Insanity” continued for many years. In 1671, another English jurist, Lord Hale, described a mentally deranged individual as “laboring under melancholy distempers.”³ In 1724, about 50 years after Lord Hale’s characterization, Lord Tracey wrote that to be exempted from punishment as a madman, a criminal defendant must be “totally deprived of his understanding and memory and doth not know what he is doing, no more than an infant, a brute or a wild beast.”³

This insanity exemption did not apply at that time across the Atlantic Ocean, in the Massachusetts Bay Colony. In that jurisdiction, in 1638, a woman named Dorothy Talbye, with “trouble of mind,” was thrown out of the church in Salem because of various actions, including assaulting her husband. She sank further into despondency and murdered her 3-year-old daughter because she said that Satan had told her to do it. The Bay Colony used the English common law that called for the death penalty in cases of murder, and Massachusetts law had no exemption based on any concept of insanity. The Bible was the ultimate source of punishment: It offered no alternatives to the death penalty. Dorothy Talbye was hanged in early 1639. Two years later, in 1641, however, the Massachusetts “Body of Liberties” was enacted, which provided that various classes of individuals (including “children, idiots, and distracted persons...”) have such allowances and dispensations in any cause “...as religion and reason require.”⁴

During those early years of English jurisprudence, attempts were made to develop standards that could be used to separate an ordinary murderer from one who is, using Justice Tracy’s word, a “madman.” But other legal jurisdictions during the Enlightenment were not as understanding or advanced in the humane and fair application of such knowledge as was England. The case of Robert François Damiens in France is an example.

*Damiens was probably mentally disturbed. He attempted a clumsy assassination of Louis XV on January 5, 1757. He was immediately arrested, and then tortured for two months by French authorities in an effort to determine his mental state at the time of the assassination attempt. Finally, on March 28, 1757, his flesh was torn away in a public square by red-hot pincers before he was drawn and quartered by six horses. This was the last such execution to occur in France.*⁶

The problem of lacking a legal standard to distinguish between insane and non-insane offenders persisted well into the 19th century. Many cases illustrate this problem; one of the most notable is that of Richard Lawrence.

*Lawrence was a relatively unknown American who thought he was King Richard III of England. On January 30, 1835, while Lawrence was apparently in a psychotic and delusional state of mind, he confronted President Andrew Jackson and fired two pistols at him. The guns failed to work properly and Jackson knocked the would-be assassin to the ground with his heavy cane. Lawrence claimed not only that he was a dead king, but that President Jackson was his clerk, with whom he had had a lengthy dispute and who he felt “deserved to be punished.” Lawrence was examined by two physicians, deemed to be insane, and institutionalized indefinitely.*⁶

The Lawrence case and its subsequent legal decision

was the first in the United States to have a significant impact on the jurisprudential problem of how to deal with alleged perpetrators of violence who were not of sound mind.

Another such case took place several years later on a London street, on January 20, 1843. On that day, a Scottish woodcutter named Daniel M’Naghten was suffering from his long and strongly held paranoid psychotic delusion that the English Prime Minister, Sir Robert Peel, wanted to kill him. He approached Peel and tried to shoot him at close range. He missed Peel and instead killed Peel’s secretary, Edward Drummond. A trial was held.

Medical testimony was produced by the defense that was successful in proving to the court that the defendant was psychotic and therefore not responsible for his actions. The acquittal created a firestorm throughout Victorian England, from the halls of Buckingham Palace to the public-at-large. In the wake of the M’Naghten decision, the British House of Lords ordered the Lords of the Queen’s Bench to create a set of legal guidelines for future cases in which the insanity defense was raised. Later, in the same year the trial was held (1843), the Queen’s Bench issued the “M’Naghten Rule,” which stated:

“...To establish a defense on the ground of insanity, it must be clearly proved that, at the time of the committing of the act, the party accused was laboring under such a defect of reason, from disease of the mind, as not to know the nature and quality of the act he was doing; or if he did know it, that he did not know (what) he was doing was wrong.”⁷

Table 1 lists M’Naghten and other variants of “Legal Insanity” in Anglo-American jurisprudence. The M’Naghten Rule was later adopted in an American jurisdiction (New Hampshire, in 1871). Nevertheless, before that, a lurid “insanity” case had occurred in Washington, DC.

On February 27, 1859, US Representative Daniel Sickles (Democrat-New York) encountered US District Attorney Philip Barton Key (the son of Francis Scott Key who wrote the poem *Defence of Fort McHenry*, from which the lyrics for The Star-Spangled Banner were taken) in broad daylight in Lafayette Park in Washington, DC, across the street from the White House. Sickles correctly believed that Key was having an affair with his wife. He took a revolver out of his pocket and shot Key, killing him instantly.

The subsequent criminal trial of Representative Sickles featured an “all-star” defense team, headed by Edwin Stanton (later President Lincoln’s Secretary of War), and based its defense on the idea that Sickles was

Table 1.

Variants of “Legal Insanity” in Anglo-American Jurisprudence	
M’Naghten test	1843
Durham Rule (“Product Test”)	1954-1972
Brawner Rule	1972
Insanity Defense Reform Act (“IDRA”)	1984
Model Penal Code (American Law Institute)	1984

suffering from temporary insanity brought on by his knowledge of his affair. An all-male jury deliberated for 2 hours and acquitted Sickles. This was the first American case in which insanity was used as a criminal defense.

After the M’Naghten Rule came to America, it was applied in a Presidential assassination case. Charles Guiteau, a frustrated office seeker and bizarre individual, was charged in 1881 with the assassination of President James Garfield. The shooting was carried out on July 2, 1881, in Union Station in Washington, DC.

In retrospect, Guiteau was probably psychotic, paranoid, and delusional. Factors of his having pre-planned the shooting underscored Guiteau’s level of premeditation of the act. An insanity defense was not offered, and Guiteau was found guilty of murder and hanged on June 30, 1882.

Although Guiteau was insane and was executed, the application of the law in cases of mental illness was not even-handed. On June 25, 1906, the clearly paranoid schizophrenic (and heir to a \$50 million fortune), Harry Thaw, shot and killed the noted architect, Sanford White, at a New York City restaurant before at least 100 witnesses. The “Trial of the Century” featured the expert testimony of some prominent “alienists” (mental health professionals, especially psychologists, at the time). These experts asserted that Thaw had suffered a “brainstorm” brought on by his belief that White had deflowered Thaw’s girlfriend, Evelyn Nesbitt, the “It-Girl” of Victorian America. After one trial in which the jury could not agree on a verdict, a second one was held in which Thaw was acquitted on the grounds of insanity. Thaw then spent several years in a mental institution, and then declared cured of his mental illness.

Other noteworthy early 20th century insanity cases included that of John Schrank. Schrank attempted to

kill President Theodore Roosevelt in 1912. Schrank was subsequently examined by physicians and committed to a mental hospital indefinitely before a trial took place. His commitment turned out to be permanent.¹⁰

For years after the M’Naghten Rule was promulgated in this country, courts had tried to modify or abandon it altogether. Other defenses were adopted, including the concept of an “irresistible impulse.” That defense, in turn, was based on the idea that an individual may have “known” (cognitively) that an act was illegal, but because of a mental impairment, he or she had lost control over his or her actions. Ohio was the first state to adopt such a defense in 1834, and in 1994, Lorena Bobbit successfully interposed that defense when charged with amputating her husband’s penis.

Probably the most serious challenge to the M’Naghten Rule occurred in 1954 when the US Court of Appeals for the District of Columbia adopted the Durham Rule, or “Product Test.” That Rule was based on the idea that “an accused is not criminally responsible if his unlawful act was the product [emphasis added] of mental disease or defect.” However, in 1972,² the same court that had created the Durham Rule rejected it and accepted the Brawner Rule. This effectively put greater faith in a jury—and less in expert testimony—to determine the question of insanity.

In more recent years, a number of cases has evoked public anger and dismay when the insanity defense has been successfully employed. The Hinckley and Yates cases are examples.

On March 30, 1981, John Hinckley, Jr.—with a history of mental illness—attempted to shoot and assassinate President Ronald Reagan. He failed to hit the President directly but wounded the President’s press secretary, James Brady, a Secret Service agent, and a police officer. The subsequent trial took place in the District of Columbia, and Federal law

therefore applied. Specifically, the Brawner Rule² contained elements of M’Naghten and the theory of irresistible impulse, and required that an individual who is suffering from a mental disease or defect is not responsible for his criminal actions if he lacks a “substantial capacity”^{*} to appreciate the criminality or to conform his conduct to the law. Hinckley’s defense successfully used this defense at trial, resulting in Hinckley’s civil commitment to a mental hospital

(St. Elizabeth’s Hospital in the District of Columbia), where he remains hospitalized to this day.

In the wake of Hinckley’s NGI verdict, the wave of public dismay—if not outright disgust—with the decision was reminiscent of that which had occurred in Great Britain after the M’Naghten decision over 100 years before. In direct reaction to the Hinckley decision, however, Congress passed the Insanity Defense Reform Act (“IDRA”) in 1984. The IDRA

Table 2. *Prominent NGI/NGRI & Related Cases*

Name	Date	Outcome
Dorothy Tallybe	1638	NGI not provided for in the Bible
Richard Lawrence	1835	NGI/NGRI ¹
Daniel M’Naghten (UK)	1843	NGI/NGRI
Daniel Sickles	1859	NGI/NGRI
John Wilkes Booth	1865	Not tried
Charles Guiteau	1882	Convicted ²
Jack the Ripper (UK)	1888	?
Harry Thaw	1906	NGI/NGRI ³
John Schrank	1912	NGI/NGRI
Leopold and Loeb	1924	Unsuccessful irresistible impulse defense
Ezra Pound	1946	Unfit to stand trial ⁴
David Berkowitz (“Son of Sam”)	1978	Convicted ⁵
John Hinckley	1982	NGI/NGRI
Lorena Bobbitt	1994	NGI/NGRI
Andrea Yates	2001	Convicted, then NGI/NGRI ⁶

1. NGI/NGRI pre-M’Naghten

2. Convicted; NGI/NGRI not considered

3. Two Trials: Hung jury, then NGI/NGRI

4. Twelve-year psychiatric hospitalization (civil commitment) at St. Elizabeth’s (Federal) Hospital, Washington, DC

5. Refused insanity plea

6. Convicted in 2002; reversed, retried, and found NGI/NGRI in 2006.

reiterates the M’Naghten standard, except that it also required that a defendant suffer from a “severe mental defect,” and it placed the burden of proving that defect or illness on the defendant by a different standard, viz., that of “clear and convincing evidence.”¹¹

Several other well-known individuals might have had, or did have, a “Legal Insanity” psychiatric defense. Although infamous and quite possibly psychotic, “Jack the Ripper” (1888) was and is sufficiently unknown to permit legal scholarship to decide whether or not he would have been considered “Legally Insane” (by the 50-year-old M’Naghten standard, at the time) of his crimes. Leopold and Loeb (1924) had been examined by psychiatrists, found legally sane by the time of trial, failed to establish an “irresistible impulse” defense at trial, and ultimately sentenced to life imprisonment for their murder of Bobby Franks. Ezra Pound, the poet and alleged quisling, was found not competent to stand trial (1946) and served a 12-year period of involuntary psychiatric hospitalization before being discharged. Although likely psychotic and possibly legally insane, David Berkowitz (“Son of Sam”; 1977) refused to permit an insanity defense to be used by his counsel and was eventually convicted of multiple murders.

Most recently—and the last in the series of cases presented in this article—is that of Andrea Yates, a young Houston, Texas housewife and mother, who drowned her 5 young children in a bathtub on June 20, 2001. Andrea Yates had had a longstanding history of mental illness, including 2 lengthy psychiatric hospitalizations prior to the drowning and a previous diagnosis of postpartum psychosis. In her first trial in 2002, an attempt to use an insanity defense (based

on the M’Naghten Rule) failed. Ms. Yates was convicted and sentenced to life in prison. It was later determined that inaccurate testimony had been given by the prosecution’s psychiatrist; the conviction was subsequently overturned, and the verdict from a new trial in 2006 was “not guilty by reason of insanity” for Ms. Yates.¹²

Table 2 summarizes the 15 cases described in this article.

Concluding Remarks: Issues, Dilemmas, and Clinical Aspects of Legal Insanity

Referring back to the variants of “Legal Insanity” tests (Table 1), many states use the M’Naghten standard and put the burden of proving mental disease or defect on the defendant. Others use the same M’Naghten standard, but place the burden of proving such a condition on the state.** Other variations among the states include those that have adopted the Model Penal Code (“MPC”) standard, and several states have adopted a law that permits a defendant to plead “guilty but mentally ill” (“GBMI”).^{13**}

Four states (Kansas, Montana, Idaho and Utah) do not allow the insanity defense. In those jurisdictions the defendant must be found competent to stand trial and then must show evidence of mental illness in order to demonstrate he or she could not have possessed the requisite intent needed for the state to show the defendant’s culpability.¹³

The debate on the issue of mental illness and its application in the various judicial systems goes on and on.

Psychiatrists, neurologists, neuropsychiatrists, psychologists, neuropsychologists, neuroimaging

**A characteristic of the “GBMI” approach is that the defendant’s fate is left to the court which can decide whether to sentence to a prison or to a mental institution, and if to the latter, how much—if any—treatment should be provided. It is possible in some cases that the adjudicated defendant will be committed to a mental hospital and then transferred to a prison to complete the balance of the imposed sentence.

specialists, biomedical ethicists and neuroethicists, other neuroscientists, as well as lawyers and judges all seek to strike the proper balance between the rights of individuals, the safety of society, and the proper application of good and ethical science to the bedeviling problem of responsibility for unacceptable behaviors of individuals who may or may not know what they did when they engaged in their behaviors.

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What's New in Neuroscience?

Sudhansu Chokroverty, MD, FRCP, FACP

This section focuses on recent articles in neuroscience that are clinically relevant for practicing neurologists, neurosurgeons and family physicians. Determining factors for selection of these articles include clinical and scientific interest, educational value and relevance in day-to-day patient care. Articles chosen for this section are derived from nationally and internationally reputable journals not widely read by practicing physicians. We hope these articles, briefly summarized and commented on, stimulate you, the reader, to look for the next issue of *JNJNI* with interest and enthusiasm.

In this issue we are directing the practicing neurologists' attention to two important topics published recently.

Dijk JM, Tijssen MA. Management of patients with myoclonus: Available therapies and the need for an evidence-based approach. Lancet Neurol 2010; 9: 1028-36.

The authors briefly reviewed drug treatment of myoclonus based on an evidence-based approach and classification of myoclonus (clinical, etiological, and anatomical location). Because of the paucity of an evidence-based approach treatment is mainly

dependent on observational studies and expert opinion. Treatment should be directed at causative agents in addition to symptomatic therapy. Treatment modalities are predominately determined by the location of the myoclonic generators (e.g., cortical, brainstem, spinal, and peripheral). The treatment is often disappointing. To develop treatment guidelines randomized, double-blind controlled drug trials are needed in the future using large homogeneous patient groups.

Dubois B, Feldman HH, Jacova C, et al. Revising the definition of Alzheimer's disease: A new lexicon. Lancet Neurol 2010; 9: 1118-27.

Based on recent advances in the use of reliable biomarkers of Alzheimer's disease (AD) giving in-vivo evidence of the disease, the authors, as part of the International Working Group, proposed new research diagnostic criteria for the AD clinical spectrum. One should consider AD as a clinical and symptomatic entity encompassing both predementia and dementia phases. We are now stepping into a new era in AD spectrum where it may be possible in the future to develop new drugs to intervene in the pathogenic cascade of the disease.

Instructions to Authors

Article Types

Original research articles and reviews should be limited to a maximum of 2000 words with 20 references, 1 table and 2 figures.

Case reports may contain up to 1000 words, 1 table and 1 figure.

What's new in neuroscience should include a brief summary and pertinent comments on recent articles in neuroscience which are clinically relevant for the practicing physicians.

Images in neuroscience articles should consist of high-resolution images (e.g., neuroimaging, polysomnographic tracing, actigraphic recording, EMG tracing, eye movement and vestibular recordings, evoked potential and EEG tracings, interesting neurosurgical specimens, etc.) derived from a specific clinical situation.

Original research articles should be organized as follows: Title page, Abstract (50 words), Introduction, Methods, Results, Discussion, References, legends, tables, figures.

Keywords of 4-6 items must be included on the title page.

Reference style should follow the Vancouver style as described in the "Uniform Requirements for Manuscripts Submitted to Biomedical Journals" (published in *N Engl J Med* 1997;336:309-315). The titles of journals should be abbreviated in conformity with Index Medicus. The following are a few examples:

1. Bondi M, Kaszniak A. *Implicit and explicit memory in Alzheimer's disease and Parkinson's disease.* *J Clin Exp Neuropsychol* 1991;13:339-358.
2. Wechsler D. *Wechsler Adult Intelligence Scale.* New York: Grune & Stratton, 1976.
3. Hirst W, Volpe B. *Automatic and effortful encoding in amnesia.* In: Gazzaniga M, editor. *Handbook of cognitive neuroscience.* New York: Plenum Press, 1984; p. 369-386.

Articles dealing with human experiments must conform to the principles enumerated in the **Helsinki Declaration** of 1975 and must include a statement that informed consent was obtained after full explanation of the procedure.

Authors must disclose any **conflicts of interest** when submitting their manuscript.

Authors must submit all figures as either **.jpeg** or **.tiff** files. Please include a legend for all figures.

Each table, figure, graph, etc., should have its relative placement noted within the text.

Papers should be **submitted electronically** (in a Word document) to the editorial office (abdrennan@solarishs.org).

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