

“THE MADNESS OF KING GEORGE”: SEVERAL SOMATOFORENSIC CASES

A Lecture for the Department of Forensic Sciences,
University of Rhode Island
Kingston, RI

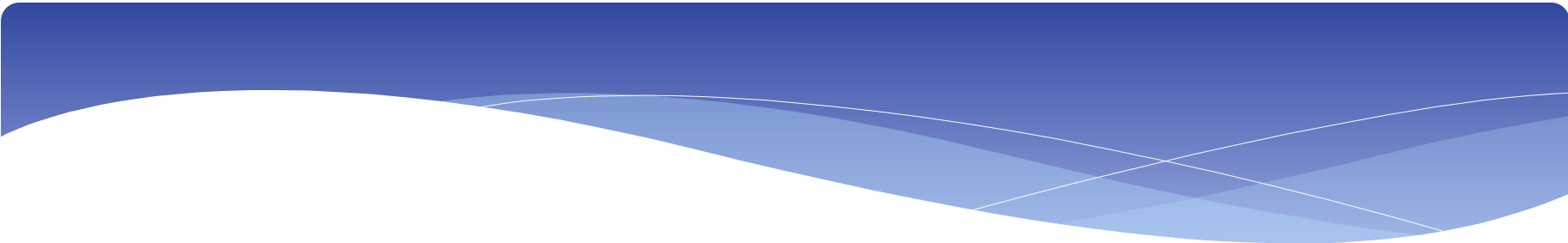
October 3, 2014

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“The Madness of King George”: Five Somatoforensic Cases

- * Definition: “SOMATO” = “SOMA (Latin), “body”
“FORENSIC” = “FORENSIS” (Latin), “of a form, place of assembly”
- * “Somatoforensic” is mainly (in my experience) used synonymously with “somatoform,” especially in the context of chronic fatigue syndrome (“CFS”) and its variants.



Overview of this Presentation

- * Introduction
- * Review of Disease Classification and Causation
(in science and in the law)
- * Review of the Five Cases and Clinical/Legal Causation
- * The Votes

Choices in the Votes

- * Guilty (or responsible) as charged
- * Not Guilty
- * Not Guilty by Reason of Insanity (“NGRI”)
- * Need more information
- * Don’t know

CAUSATION AND ASSOCIATION: I

“When we can produce a phenomenon artificially...and observe it in circumstances...with which we are accurately acquainted...,we may produce...variations to any extent, and of such kinds as we think best calculated to bring the laws of the phenomenon into clear light.”

*J.S. Mills, 1876
A System of Logic*

“Correlation is not causation; association is not causation...”

Anon. 1980

CAUSATION AND ASSOCIATION: II

Broadly speaking, a series of reported statistical association can be explained as:

- ❖ Artifactual (spurious)
- ❖ Indirect, or
- ❖ Causal or etiological

TYPES OF ASSOCIATION*

- I. NOT STATISTICALLY ASSOCIATED (INDEPENDENT)
- II. STATISTICALLY ASSOCIATED
 - * Noncausally associated (secondary association)
 - * Causally associated
 - a) Indirectly associated
 - b) Causally associated

**FROM: MACMAHON, B. AND T.E. PUGH,
EPIDEMIOLOGY PRINCIPLES AND METHODS (1970),
BOSTON; LITTLE BROWN & CO. (PAGE 18)*

PRE-GERM THEORY CONCEPTS OF DISEASE CAUSATION

- * IMBALANCE OF THE FOUR HUMORS (BLACK BILE; MELANCHOLIC; YELLOW BILE; CHOLERIC; BLOOD; SANGUINE; PHLEGM; PHLEGOMATIC)
- * RELIGIOUS BASES: CURSES, WITCHES (SPRENGER AND KRAEMER'S MALLEUS MALLIFICARUM, THE HAMMER AGAINST WITCHES)
- * MIASMA (“VAPORS”)
- * TRAUMA (PHYSICAL: HOMER'S EGYPTIAN MEDICINE)

“PROOF” OF “CAUSATION” IN MEDICINE: KOCH’S POSTULATES*

- * “THE MICROORGANISM MUST REGULARLY BE ISOLATED FROM CASES OF THE ILLNESS”;
- * “IT MUST BE GROWN IN PURE CULTURE IN VITRO”;
- * “WHEN SUCH A PURE CULTURE IS INOCULATED INTO SUSCEPTIBLE ANIMAL SPECIES, THE TYPICAL DISEASE MUST RESULT”; AND
- * “FROM SUCH EXPERIMENTALLY INDUCED DISEASE, THE MICROORGANISM MUST AGAIN BE ISOLATED”

*FROM: JAWETZ, E., J. MELNICK, AND E. ADELBERG,
REVIEW OF MEDICAL MICROBIOLOGY (SIXTH EDITION; 1984)
LOS ALTOS: LANGE MEDICAL PUBLICATIONS, PAGE 134.

THE BRADFORD HILL CAUSALITY CRITERIA*

- * STRENGTH (OF ASSOCIATION)
- * CONSISTENCY (AMONG SAMPLES AND STUDIES)
- * SPECIFICITY (OF ASSOCIATION)
- * TEMPORALITY (BETWEEN CAUSE, THEN EFFECT)
- * BIOLOGICAL GRADIENT (BETWEEN EXPOSURE AND EFFECT); DOSE RESPONSE RELATIONSHIP
- * PLAUSIBILITY (OF THE MECHANISM LINKING EFFECT WITH CAUSE)
- * COHERENCE (BETWEEN EPIDEMIOLOGIC AND LABORATORY FINDINGS)
- * EXPERIMENT (EXPERIMENTAL PROOF)
- * ANALOGY (WITH OTHER SIMILAR FACTORS)

*HILL, A.B. "THE ENVIRONMENT AND DISEASE: ASSOCIATION OR CAUSATION?"
PROC ROYAL SOC. MED., 58: 295-300, 1965

“CAUSATION” IN MEDICINE TURNED ON ITS HEAD: SAMUEL BUTLER’S EREWHON*



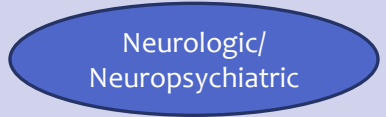

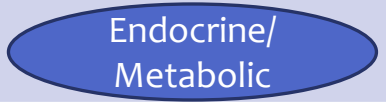


In 1872, the English author, Samuel Butler, described in a nineteenth century Utopian novel a fictitious country in which “... If a man forges a cheque, or sets his house on fire, or robs with violence from the person... he is taken to a hospital and most carefully tended at public expense (page 79) but also where disease is a crime punishable by imprisonment...” (page 242). The name of this land is “EREWHON,” (close to) “NOWHERE” in reverse.

**Butler, S. EREWHON (1872, reprinted in 1960).
New American Library*

BASES OF DISEASE CLASSIFICATION

BY ETIOLOGY	BY ORGAN SYSTEM	BY MEDICAL SPECIALTY	BY AGE	COMBINATION AND OVERLAPS
Neoplastic	Dermatologic	Palliative Care	Prenatal and Neonatal	e.g. Huntington's Disease (genetic and degenerative)
Degenerative	Hematologic	Psychiatric	Pediatrics	
Genetic	Neurologic/ Neuropsychiatric	ENT	Adolescent Medicine	
Congenital	Endocrine/ Metabolic	Surgery and Subspecialties	Internal Medicine/ Primary Care	
Physicochemical	Gastrointestinal/ hepatic	Nanomedicine	Gerontology	
Infectious	Genitourinary/Renal	PM & R		
Traumatic	Cardiovascular	Radiology/ Imaging		
Others [Idiopathic]	Pulmonary	Addiction Medicine		

THESE FIVE CASES. I

BY ETIOLOGY	BY ORGAN SYSTEM	BY MEDICAL SPECIALTY	BY AGE	COMBINATION AND OVERLAPS
Neoplastic  	Dermatologic 	_____ Care	Prenatal and Neonatal	e.g. Huntington's Disease (genetic and degenerative)
	Hematologic	Psychiatric	Pediatrics	
		ENT	Adolescent Medicine	
Congenital		Surgery and Subspecialties	Gerontology	
Physicochemical	Gastrointestinal/ hepatic			
		Nanomedicine		
Traumatic	Cardiovascular	PM & R		
Others	Pulmonary	Radiology/		

THESE FIVE CASES. II

- * INFECTIOUS/NEUROPSYCHIATRIC
(NEUROSYPHILIS)
- * ENDOCRINE/METABOLIC
(ACUTE INTERMITTENT PORPHYRIA; “AIP”)
- * DEGENERATIVE/GENETIC/NEUROLOGIC
(HUNTINGTON’S)
- * GENITOURINARY/RENAL/IDIOPATHIC
(ACUTE RENAL FAILURE, “ARF”)
- * NEUROLOGIC/IDIOPATHIC
(EPILEPSY)

THESE FIVE CASES. III

For somatoforensic cases, is it necessary to define clearly – to “carve nature at her joints”* -- the nature and pathogenesis of the disease of forensic interest?

**With apologies to Plato.*

THESE FIVE CASES. IV

NO. WHAT MATTERS IS THE SYMPTOMATOLOGY
AND/OR DISABILITY, NOT JUST THE DIAGNOSIS.

THESE FIVE CASES. V

THE “COMPLEAT” PRACTICING FORENSIC PSYCHIATRIST IS, FIRST AND FOREMOST, A PHYSICIAN AND CLINICIAN, KNOWLEDGEABLE AND ADEPT IN GENERAL MEDICINE AS WELL AS IN PSYCHIATRY, NEUROPSYCHIATRY AND THE ADDICTIONS

TIME FRAMES FOR CRIMINAL FORENSIC PSYCHIATRIC EVALUATIONS

Past	Present	Future
<i>At time of investigation</i>	<i>Competency to stand trial/proceed to trial</i>	<i>Competency to stand trial/proceed to trial</i> <i>Dangerousness</i>
<input type="checkbox"/> Legal insanity	Civil commitment (present dangerousness)	<input type="checkbox"/> Civil commitment (involuntary psychiatric hospitalization)
<input type="checkbox"/> Diminished capacity	<input type="checkbox"/> Sex offenders	<input type="checkbox"/> Sexual civil commitment (sexually violent predator [SVP]/sexually dangerous person [SDP])
<input type="checkbox"/> Intoxication	<input type="checkbox"/> Civil commitment (involuntary hospitalization)	<input type="checkbox"/> Outpatient registration of sex offenders ("Megan's Law")
<input type="checkbox"/> Other committees		<input type="checkbox"/> Outpatient civil commitment ("Kendra's Law" in New York)
<i>At time of offense</i>		
<input type="checkbox"/> <i>Miranda</i> rights waiver		

CLINICAL FORENSIC ANALYSIS*

For the clinical practitioner, the conceptual framework is some variation of identification, chief complaint, history of the present illness, pertinent past history, laboratory test data, differential diagnosis, medical diagnostic impression. For the forensic psychiatric practitioner, the four-step conceptual framework is issue, legal criteria, relevant data, and reasoning process:

1. *Issue*: What is the specific psychiatric-legal issue to be considered?
2. *Legal criteria*: In the jurisdiction in which this specific psychiatric-legal issue must be resolved, what are the legally defined terms and criteria that will be used for its resolution?
3. *Relevant data*: Exactly what information (such as part of what might be collected by a clinician following the traditional clinical framework for data organization) is there that is specifically pertinent to the legal criteria that will be used to resolve the specific psychiatric-legal issue?
4. *Reasoning process*: How can the available relevant data be applied to the legal criteria so as to yield a rationally convincing psychiatric-legal opinion?

*R. Rosner (2003)

PSYCHIATRIC-LEGAL REASONING: A THREE-STEP PROCESS*

1. Assertion of a law or law-like proposition
2. Assertion of a factual proposition
3. A deductive inference from 1 and 2

PSYCHIATRIC-LEGAL REASONING: A THREE-STEP PROCESS*

FIRST EXAMPLE:

1. Humans are the only rational bipedal animals.
2. Socrates is a rational bipedal animal.
3. Therefore, Socrates is human.

SECOND EXAMPLE:

1. Persons who are competent to stand trial have the capacity to understand the charges against them, the capacity to understand the nature of the court proceedings against them, and the capacity to cooperate with an attorney in their own defense.
2. John Doe has the capacity to understand the charges against him, the capacity to understand the nature of the court proceedings against him, and the capacity to cooperate with his attorney in his own defense.
3. Therefore, John Doe is competent to stand trial.

FAULTY PSYCHIATRIC-LEGAL REASONING: I*

“If the first premise is wrong—that is, if the legal criteria used are incorrect—then the opinion is unsupported logically.

If the second premise is wrong—that is, the available data are not relevant to the legal criteria—then the opinion is unsupported, logically...”

If the two premises are correct, the deductive inference may be wrong.

THIRD EXAMPLE:

1. All humans are rational bipedal animals
2. Socrates is a rational bipedal animal
3. Therefore, Socrates like chocolate

The fallacy is obvious.

*Rosner, R. (2005)

FAULTY PSYCHIATRIC-LEGAL REASONING: II*

FOURTH EXAMPLE:

1. People with borderline personality disorder are characteristically impulsive and aggressive.
2. Roger has been diagnosed with borderline personality disorder.
3. Therefore, Roger is not criminally responsible for slaying his wife.

What is the fallacy?

-OR-

FIFTH EXAMPLE:

1. Persons who are capable of understanding the charges against them, capable of understanding the nature of the court proceedings against them, and capable of cooperating in their own defense are competent to stand trial.
2. Richard Roe understands the charges against him, understands the court proceedings against him, and is able to cooperate in his own defense.
3. Therefore, Richard Roe was legally sane (and legally responsible) at the time when he committed the offense.

What is the fallacy?

*Rosner, R. (2005)

FAULTY PSYCHIATRIC-LEGAL REASONING: III*

SIXTH EXAMPLE:

1. “Babies are illogical.
2. Nobody is despised who can manage a crocodile.
3. Illogical persons are despised.

Answer: Babies cannot manage crocodiles.”

SEVENTH EXAMPLE:

1. “Nobody who really appreciates Beethoven fails to keep silence while the ‘Moonlight Sonata’ is being played.
2. Guinea pigs are hopelessly ignorant of music.
3. No one who is hopelessly ignorant of music ever keeps silence while the ‘Moonlight Sonata’ is being played.

Answer: Guinea pigs never really appreciate Beethoven.”

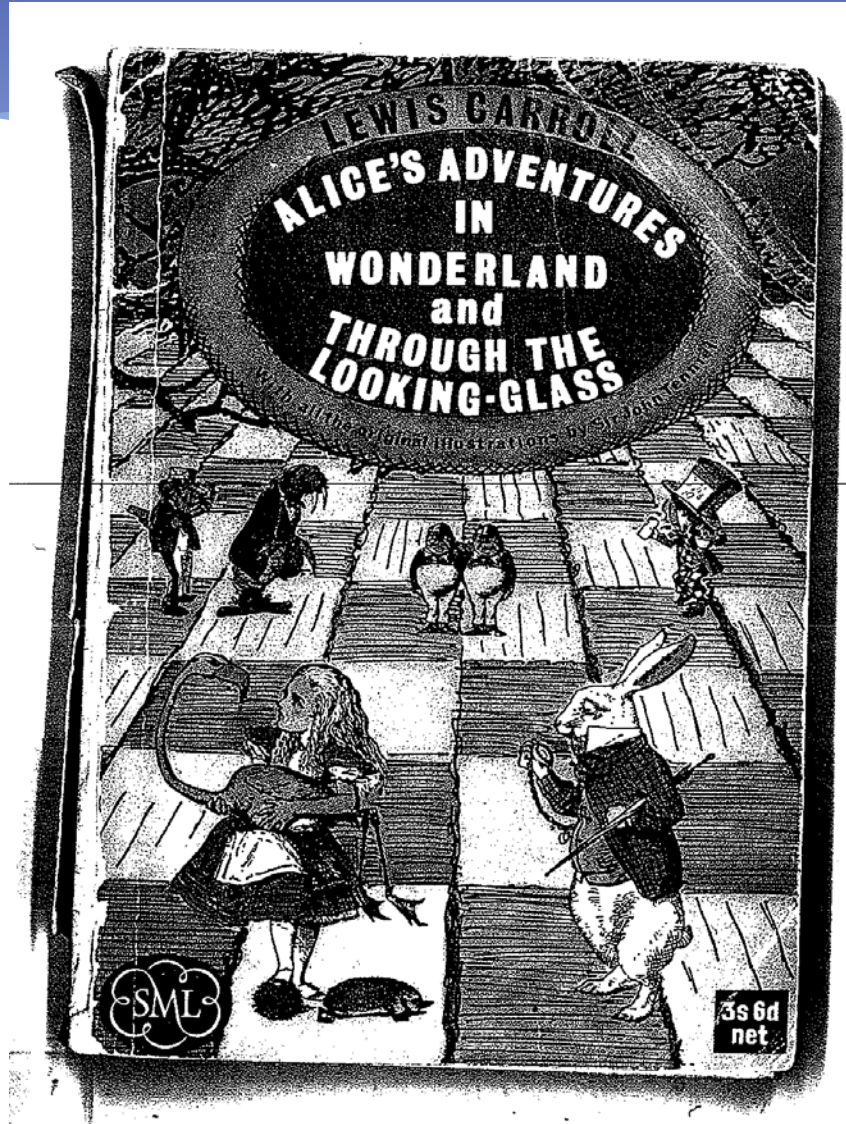
**Lewis Carroll, Symbolic Logic: Part I. Elementary (1896)*



“Drink me.”



“Smoke me.”



Clinical Decision Making

For the clinical practitioner, the conceptual framework is some variation of identification, chief complaint, history of the present illness, pertinent past history, laboratory test data, differential diagnosis, medical diagnostic impression, and treatment plan.

PSYCHIATRIC-LEGAL (FORENSIC) DECISION MAKING

For the forensic psychiatric practitioner, the four-step conceptual framework is issue, legal criteria, relevant data, and reasoning process.

PSYCHIATRIC DEFENSES TO CRIMINAL ACTS IN NEW JERSEY: THREE SPECIFIC DEFENSES*

- * Insanity Defense (2C:4-1)*
- * “Diminished Capacity” Defense (2C:4-2)*
- * Intoxication (involuntary, generally) Defense (2C:4-8)*

These are potential exceptions to the often-heard adage,

“If you do the crime, you’d better be ready to the time...”

— “Baretta” television series, 1970’S

**N.J. Code of Criminal Justice
(various editions)*

PSYCHIATRIC DEFENSES TO CRIMINAL ACTS IN NEW JERSEY: INSANITY DEFENSE

2C:4-1. Insanity Defense

A person is not criminally responsible for conduct if at the time of such conduct he 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. Insanity is an affirmative defense which must be proved by a preponderance of the evidence. L.1978, c.95

Source: NJS 2A:163-2; 2A:163-3

*Prior Laws: R.S. 2:190-171 (L. 1943, c.41. p.81, § 1)L.1922, c. 101 § 3
p. 189 [1924 Suppl. § 53-133p]*

AN HISTORICAL DIGRESSION: I

REX V. M'NAGHTEN (1843)

In the landmark *Rex v. M'Naghten* case in 1843 – which is the current basis for the insanity defense in most jurisdictions in the United States and the United Kingdom – Daniel M'Naghten, while in a delusional mental state, mistakenly shot and killed Edward Drummond, the private secretary to Sir Robert Peel, the English Prime Minister at the time, believing that he was the Prime Minister.

AN HISTORICAL DIGRESSION: II

REX V. M'NAGHTEN (1843)

Nine psychiatrists testified as expert witnesses to M'Naghten's mental state at the time of the shooting. He was found legally insane, even though testimony indicated that he might have generally been able to conduct his life rationally and have been able to understand the difference between right and wrong.

AN HISTORICAL DIGRESSION: III

REX V. M'NAGHTEN (1843)

The amorphous quality of M'Naghten's mental condition described by the experts in this case which permitted a successful insanity defense left the Victorian crown, government, and public uncertain. This uncertainty resulted in a subsequent ruling by a commission of fifteen Queen's Bench judges giving the following well-known language – language which in psychiatric “legal insanity” defense in English-based legal jurisdictions to this day – for that defense.

AN HISTORICAL DIGRESSION: IV

REX V. M'NAGHTEN (1843)

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

PSYCHIATRIC DEFENSES TO CRIMINAL ACTS IN NEW JERSEY: “DIMINISHED CAPACITY” DEFENSE

2C:4-2. Evidence of mental disease or defect admissible when relevant to element of the offense.

Evidence that the defendant suffered from a mental disease or defect is admissible whenever it is relevant to prove that the defendant did not have a state of mind which is an element of the offense. In the absence of such evidence, it may be presumed that the defendant had no mental disease or defect which would negate a state of mind which is an element of the offense. Mental disease or defect is an affirmative defense which must be proved by a preponderance of the evidence. L.1978 c.

The Four Levels of Complicatedness of Behavior (“Conduct”) Articulated in the “Diminished Capacity” Psychiatric Criminal Defense Statute

1. For the first- and second-degree offenses (more serious; more punishment, if convicted)
 - * Purposeful
 - * Knowing
2. For third- and fourth-degree offenses (less serious; less punishment, if convicted)
 - * Reckless
 - * Negligent

PSYCHIATRIC DEFENSES TO CRIMINAL ACTS IN NEW JERSEY: INTOXICATION DEFENSE

2C:2-8. Intoxication

- a. Except as provided in subsection d. of this section, intoxication of the actor is not a defense unless it negatives an element of the offense.
- b. When recklessness establishes an element of the offense, if the actor, due to self-induced intoxication, is unaware of a risk of which he would have been aware had he been sober, such unawareness is immaterial.
- c. Intoxication does not, in itself, constitute mental disease within the meaning of chapter 4.
- d. Intoxication which (1) is not self-induced or (2) is pathological is an affirmative defense if by reason of such intoxication the actor at the time of his conduct did not 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. Intoxication under this subsection must be proved by clear and convincing evidence.

PSYCHIATRIC DEFENSES TO CRIMINAL ACTS IN NEW JERSEY: INTOXICATION DEFENSE

2C:2-8. Intoxication – cont.

- e) Definitions. In this section unless a different meaning plainly is required:
- 1) “Intoxication” means a disturbance of mental or physical capacities resulting from the introduction of substances into the body;
 - 2) “Self-induced intoxication” means intoxication caused by substances which the actor knowingly introduces into his body, the tendency of which to cause intoxication he knows or ought to know, unless he introduces them pursuant to medical advice or under such circumstances as would afford a defense to a charge of crime;
 - 3) “Pathological intoxication” means intoxication grossly excessive in degree, given the amount of the intoxicant, to which the actor does not know he is susceptible. *L.1978, c. 95, § 2C:2-8, eff. Sept. 1, 1979. Amended by L.1983,c. 306. § 1, eff. Aug. 26, 1983.*

Source: Model Penal Code: 2.08.

TIME FRAMES FOR CRIMINAL FORENSIC PSYCHIATRIC EVALUATIONS

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COMPETENCY TO STAND TRIAL IN NEW JERSEY: “MENTAL INCOMPETENCE EXCLUDING FITNESS TO PROCEED”

2C:4-4. Mental incompetence excluding fitness to proceed.

- a. No person who lacks capacity to understand the proceedings against him or to assist in his own defense shall be tried, convicted or sentenced for the commission of an offense so long as incapacity endures.
- b. A person shall be considered mentally competent to stand trial on criminal charges if the proofs shall establish:
 - 1) That the defendant has the mental capacity to appreciate his presence in relation to time, place and things; and
 - 2) That his elementary mental processes are such that he comprehends
 - a. That he is in a court of justice charged with a criminal offense.
 - b. That there is a judge on the bench.
 - c. That there is a prosecutor present who will try to convict him of a criminal charge.
 - d. That he has a lawyer who will undertake to defend him against the charge.
 - e. That he will be expected to tell to the best of his mental ability the facts surrounding him at the time and place where the alleged violation was committed if he chooses to testify and understands the right not to testify.
 - f. That there is or may a jury present to pass upon evidence adducted as to guilt or innocence of such charge or that if he should choose to enter into plea negotiations or to plead guilty that he comprehend the consequences of a guilty plea and that he be able to knowingly, intelligently, and voluntarily waive those rights which are waived upon such entry of a guilty plea. And
 - g. That he has the ability to participate in an adequate presentation of his defense.
- 3) *L.1978, c. 95, § 2C:4-4, eff. Sept. 1, 1979. Amended by L.1979, c. 178. § 13, eff. Sept. 1, 1979.*

Source: N.J.S., 2A 163-02
Model Penal Code: 4.04.

CASE PRESENTATION

- * Middle-class, working adult
- * Several months before incident – complaint of visual loss
- * Dx – neurosyphilis (psychotic-level)
- * Day of incident – delusional behavior/illegal actions (fire-setting incident)
- * Apprehended/arrested
- * Hospitalized – vision continued to decline
- * ID consult
- * Dx – neurosyphilis
- * Temporarily placed in convalescent center
- * Returned home
- * Trial

SYPHILIS

“The Great imitator...”

“He who knows syphilis know medicine.”

Sir William Osler

SYPHILIS

“THE DISEASE OF THREE’S”

3 stages

3 weeks 3 months 3 years

3 primary systems

- * Gumma/CV/Neuro

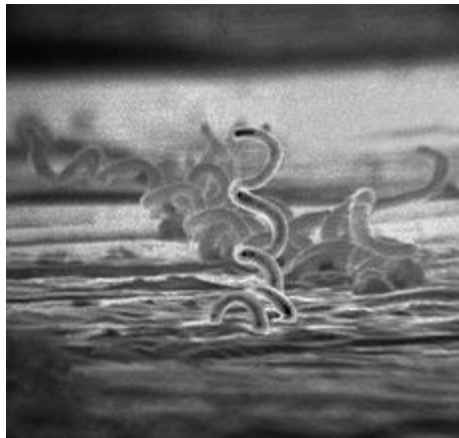
3 treatment courses

- * IM x 1, IM x 3, IV

THE STATES AND NATURAL HISTORY OF SYPHILIS

- 1) Primary State (“Chancre” – 3-6 weeks)
- 2) Secondary State (rash, adenopathy, fever, lethargy, malaise, sore throat, headache – 2-6 weeks)
 - * Early latent syphilis (up to one year post-infection)
 - * Late latent syphilis (> one year post-infection)
 (“neurosyphilis” manifesting 20-30 years post-infection; ca. 10% of untreated cases)
- 3) Tertiary (Late) Stage (ongoing inflammatory disease, affecting the aorta, lungs, eyes, and other organs, through gumma formation; ca. 10% of untreated cases)

CAUSATIVE ORGANISM OF SYPHILIS



Electron micrograph of *Treponema pallidum*

PRIMARY SYPHILIS



Primary chancre of syphilis on the hand

SECONDARY SYPHILIS



Reddish papules and nodules over much of the body due to secondary syphilis



The earliest known medical illustration of patients suffering from syphilis, Vienna, 1498

FOUR VARIANTS OF TERTIARY (LATE) NEUROSYPHILIS

1. Asymptomatic neurosyphilis (non-specific symptomatology; headaches, confusion)
2. Meningovascular syphilis (vascular occlusion and CVA's, 6-7 years after the initial infection)
3. Parenchymal Neurosyphilis
 - * Tabes dorsalis (peripheral nervous system)
4. Parenchymal Neurosyphilis
 - * General paresis (general paresis of the insane, or "GPI"; "dementia paralytica" of the central nervous system)

PARENCHYMAL NEUROSYPHILIS: GPI: DEMENTIA-PARALYTICA ("P.A.R.E.S.I.S.")

Paresis (personality changes, psychotic behaviors, psychotic delusions)

Affect (grandiose, delusional, "insane," mania)

Reflexes (hyperreflexia)

Eye (Argyll Robertson pupil, visual disturbances)

Sensorium (hallucinations, illusions)

Intellect (memory loss, poor focus and concentration, impaired judgment and insight, other cognitive dysfunctions)

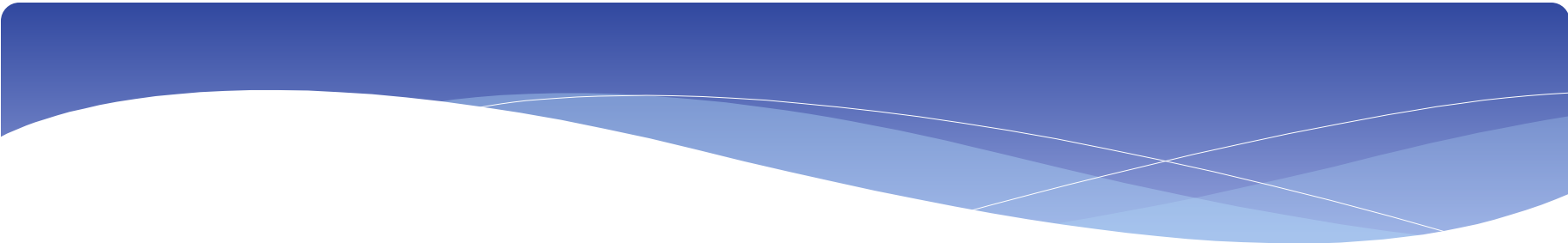
Speech (speech disturbance)

CLINICAL DECISION MAKING

For the clinical practitioner, the conceptual framework is some variation of identification, chief complaint, history of the present illness, pertinent past history, laboratory test data, differential diagnosis, medical diagnostic impression, and treatment plan.

CASE PRESENTATION

- * Young mother of a 2-year-old, single parent, employed.
- * Several-year history of unexpected acute abdominal pain, nausea, and vomiting, lasting hours-to-days, unpredictable, accompanied by agitation, anxiety, and periodically frank psychosis, and evaluated in multiple E.D. visits.
- * Typically has “warning” at onset of episode, and arranges for care of her child, with relatives.
- * Child Protective Services has instituted proceedings to terminate her parental rights.
- * Somatoforensic evaluation includes Acute Intermittent Porphyria (“A.I.P.”) in her differential diagnosis.
- * Subsequent laboratory and clinical evaluations confirm “a porphyria” but not specifically A.I.P.
- * A hearing was held to address termination of parental rights, at which the somatoforensic evaluator testified.



ACUTE INTERMITTENT PORPHYRIA

“...Though there are several different types of porphyrias, the one with the most serious consequences and the one that usually presents in adulthood is acute intermittent porphyria, which is inherited as an autosomal dominant, though it remains clinically silent in the majority of patients. Those who develop clinical illness are usually women, with symptoms beginning in the teens or 20’s... The disorder is caused by partial deficiency of porphobilinogen deaminase activity, leading to increased excretion of amino leveling acid and porphobilinogen in the urine. The diagnosis may be elusive if not specifically considered [emphasis added]...characteristic abdominal pain...Attacks are precipitated by numerous factors, including drugs and intercurrent infections...”

--R.E. Pyeritz, in *2006 Current Medical Diagnosis and Treatment*
(45th Edition)

TREATMENT OF ACUTE INTERMITTENT PORPHYRIA*

* Prevention

- Avoid sulfonamides, barbiturates, and drugs listed in Table
- Avoid starvation diets

* Treatment of attacks

- High-carbohydrate diet (long-term)
- Analgesics and antipsychotics (acute)
- Intravenous glucose and hematin (controversial) (acute)
- Liver transplantation (last ditch)

**After R.E. Pyeritz (2006)*

Table 41-1. Some of the “unsafe” and “probably safe” drugs used in the treatment of acute porphyrias

Unsafe		Probably Safe	
Alcohol	Meprobamate	Acetaminophen	Imipramine
Alkylating Agents	Methyldopa	β-Adrenergic blockers	Insulin
Barbiturates	Metoclopramide	Amitriptyline	Lithium
Carbamazepine	Nortriptyline	Aspirin	Naproxen
Chloroquine	Pentazocine	Atropine	Nitrofurantoin
Chlorpropamide	Phenytoin	Chloral hydrate	Opioid analgesics
Clonidine	Progestins	Chlordiazepoxide	Penicillamine
Dapsone	Pyrazinamide	Diazepam	Penicillin and derivatives
Ergots	Rifampin	Digoxin	Phenothiazines
Erythromycin	Spirolactone	Diphenhydramine	Procaine
Estrogens, synthetic	Succinimides	Glucocorticoids	Streptomycin
Food additives	Sulfonamides	Guanethidine	Succinylcholine
Glutethimide	Theophylline	Hyoscine	Tetracycline
Griseofulvin	Tolazamide	Ibuprofen	Thiouracil
Hydralazine	Tolbutamide		
Ketamine	Valproic acid		

LabCorp
Laboratory Corporation of America

LabCorp
NJ Avenue

Phone: 800

Patient Name					Specimen Number		
Account Number	Patient ID	Control Number	Date and Time Collected	Date Reported	Sex	Age(Y/M/D)	Date of Birth
			09/ / 09:53	09/ / /			

TESTS	RESULT	FLAG	UNITS	REFERENCE INTERVAL	LAM
Hexaporphyrin	>1.0 ug/dL	Elevated			
Reference Range	<1.0		ug/dL	0.0-1.0	02
< or = 1.0 ug/dL	Normal				
Pentaporphyrin	>1.0 ug/dL	Elevated			
Reference Range	<1.0		ug/dL	0.0-1.0	02
< or = 1.0 ug/dL	Normal				
Protoporphyrin	>1.0 ug/dL	Elevated			
Reference Range	<1.0		ug/dL	0.0-1.0	02
< or = 1.0 ug/dL	Normal				
Coproporphyrin	>1.0 ug/dL	Elevated			
Reference Range	<1.0		ug/dL	0.0-1.0	02
< or = 1.0 ug/dL	Normal				

The performance characteristics of the listed assay was validated by Cambridge Biomedical Inc. The US FDA has not approved or cleared this test. The results of these assay can be used for clinical diagnosis without FDA approval. Cambridge Biomedical is a CLIA certified, CAP accredited laboratory for performing high complexity assays such as this one.

Porphyrins, Qn, Random U

Uroporphyrins (UP)	2		ug/L	0 - 20	03
Heptacarboxyl (7-CP)	<1		ug/L	0 - 2	03
Hexacarboxyl (6-CP)	3	High	ug/L	0 - 1	03
Pentacarboxyl (5-CP)	1		ug/L	0 - 2	03
Coproporphyrin (CP) I	43	High	ug/L	0 - 15	03
Coproporphyrin (CP) III	34		ug/L	0 - 49	03

Folate (Folic Acid), Serum

Folate (Folic Acid), Serum	14.5		ng/mL	>3.0	01
Note:					01

A serum folate concentration of less than 3.1 ng/mL is considered to represent clinical deficiency.

CASE PRESENTATION

- * A 42-year-old male with a Family History of Huntington's Disease allegedly assaulted a neighbor while "confused," thinking that the neighbor was going to "rob me." He had a 20-year history of less serious offenses and arrests, 3 directed against the same neighbor over the past four years.
- * A defense ("diminished capacity") was raised by defense counsel for the most recent offense, based on the possibility of a diagnosis of Huntington's Disease.
- * Subsequent clinical and laboratory evaluation of the male was consistent with Huntington's Disease.
- * Defense counsel arranged for a somatoforensic evaluation of the male, taking into account his history, his behaviors surrounding the time of the offense, and his recent genetic test results for Huntington's Disease.
- * The somatoforensic evaluator presented his observations, findings, impressions, and expert opinions to defense counsel.

Huntington's Disease

* DEFINITION

- Inherited progressive neurodegenerative disorder
- Characterized by choreiform movements, psychiatric problems and dementia
- Caused by trinucleotide (CAG) expansion in the Huntington gene located on Chromosome 4p16.3

Genetics of Huntington's Disease

* GENETICS

- Autosomal dominant inheritance
- More than 35 trinucleotide repeats results in an unstable, disease causing allele.
 - Adult form of HD → 40-50 CAG repeats
 - Juvenile form of HD → greater than 60 CAG repeats
- Genetic Anticipation

Pathophysiology of Huntington's Disease

* PATHOPHYSIOLOGY

- Aggregation of mutant Huntingtin is a pathologic hallmark of the disease process
- Neuronal Loss
- Marked atrophy of the neostriatum

MRI Coronal Section of Huntington's Disease



Coronal section from a MR brain scan of a patient with HD showing atrophy of the heads of the caudate nuclei, enlargement of the frontal horns of the lateral ventricles (hydrocephalus ex vacuo), and generalized cortical atrophy.

Clinical Progression of Huntington's Disease

- ❖ Insidious onset of symptoms with movement abnormalities or psychiatric symptoms.
 - Chorea
 - Hypotonia with hyperreflexia
 - Motor impersistence
- ❖ As disease progresses, chorea gradually is replaced by a parkinsonian akinetic rigid state
- ❖ Abnormal eye movements

Psychiatric Symptoms in Huntington's Disease

- * Psychiatric symptoms

- Symptoms do not correlate with duration of the disease, repeat length, or presence of dementia or motor symptoms
- Early onset → Irritability, depression, disrupted relationships
- Depression, paranoia, delusions and hallucinations can develop at any point in the illness
- HD is associated with increased risk of suicide for diagnosed patients, at risk family members and even those shown to be negative for the mutation

Cognitive/Neuropsychiatric Features in Huntington's Disease

* Cognitive Symptoms

- Executive dysfunction
- Lack of insight into their cognitive symptoms
- Difficulty with time based tasks
- Have greater improvement with cuing during recall tasks
- Memory loss is usually a late finding
- Aphasia and apraxia are uncommon

CASE PRESENTATION: I

- * A.B., a 68 year-old divorced white male of eastern European origins, had immigrated to the U.S. in 1990, staying with family members.
- * He worked as a self-employed truck driver for many years, maintaining his own business, which he described as “the reason for all of my [chronic] headaches,” and which he self-medicated with high doses of aspirin on an ongoing basis.
- * A.B. described himself as a “social drinker” and not a drug user. Medically, he is overweight, has Type II Diabetes Mellitus, is moderately hypertensive, and is generally noncompliant/non-adherent with his medications

CASE PRESENTATION: II

- * A.B. became vaguely aware early one morning that his former brother-in-law in Eastern Europe had “problems with [my] business, and needs to see me right away.” He believes he heard his brother-in-law’s voice “speaking to me,” and tried unsuccessfully to Skype him.
- * A.B. then bought an airplane ticket to Eastern Europe, driving 5 hours to an international airport to leave on his flight. He left his “rig” (truck) double-parked.
- * A.B. was told that the ticket he had purchased was for a flight that was due to leave in two hours from a different, distant airport. He bought a new ticket for a flight to a different city leaving from the airport where he was. He did not have a visa for the country where his brother-in-law lived; he was vaguely worried about that.

CASE PRESENTATION: III

- * A.B. succeeded in contacting his brother-in-law, who eventually met him at an airport distant from him, and shepherded A.B. through customs and security. By this time, A.B. had been travelling about 20 hours.
- * A.B. left for return to the U.S. after a brief visit with his former brother-in-law, who was very puzzled by A.B.'s story and behavior.
- * A.B. landed in the U.S. after an 8-hour flight, having slept fitfully on the flight, and landing at the same airport from which he had departed. He found his truck, and managed to try to drive it. He drove it to a 7-11 convenience store about five miles from where he lived, to "...buy coffee and more aspirins. My head was killing me..."

CASE PRESENTATION: IV

- * When A.B. tried to use his truck, it didn't start. Instead, he got into a car with a running engine in the 7-11 parking lot and drove off, uncertain about where he was and where he wanted to go. By this time, A.B. had been travelling about three days.
- * Police chased A.B. in his stolen car. He was frightened and convinced that they were agents from the Eastern Europe country he had just visited, sent to assassinate him for not having a visa. He drove over 100 miles an hour for fifteen miles, to elude them.

CASE PRESENTATION: V

- * Toward the end of the chase, A.B. crashed into another vehicle, was apprehended by police, and had a fight with the arresting officer. The officer noted that he seemed “... wild-eyed and crazy.”
- * A.B. was then taken to a County Jail, then to a local hospital E.D., where his confused and psychotic mental state was noted and where his considerable lower extremity edema was also noted. Further examination and laboratory testing were done, and A.B. was then admitted to the hospital for further work-up.

Note to Police: I

Plis HELP MI

I. WOS POIZEN I NID OLOKTOR

Note to Police: II

“PLEASE HELP ME
I WAS POISONED. I NEED DOCTOR”

Laboratory Data

Aug Page of

Medical Center
Chart Review Print

Location	Patient Name	Patient MRN	Visit #	Age	Sex
Date of Birth	Attending Physician	Admit Date	Discharge Date		
Feb		May	Jun		

Discharge Summary LansangM
Status: complete

Dictated By:
Report:
DATE OF ADMISSION: May
DATE OF DISCHARGE: June
DISCHARGE DIAGNOSES:

1. Severe rhabdomyolysis, resolving.
2. Acute renal failure secondary to #1; dialysis required on multiple occasions. Dialysis discontinued as of June
3. Psychosis, resolved.
4. Right calf pain, resolved.
5. Hypertension.
6. Hypercholesterolemia.
7. Mental status changes, resolved.
8. Hyperkalemia.
9. Encephalopathy.
10. Leukocytosis.
11. Acute respiratory failure, requiring bilevel positive airway pressure (BiPAP).
12. Probable sleep apnea.

REASON FOR ADMISSION:

For full details regarding the admission, please see admission note.
The patient is a -year-old gentleman brought in from County Jail because of mental status changes. According to the records from prison, the patient is from but has lived here 30 years. He was arrested for stealing a car and resisting arrest. He became quite paranoid and felt as if he was being poisoned in the jail and, according to one of the officers, was wiping his mouth with a towel until he induced bleeding. He was spitting at the officers and becoming increasingly combative and they restrained him. At that juncture he was brought to the emergency room. He has not been eating because of paranoia and a fear of being poisoned.

In the emergency department he had a lumbar puncture and head computed tomography (CT scan), both of which were negative. Because of agitation he was given Ativan, Haldol, and Cogentin. He also received

Page of

Medical Center
Chart Review Print

Location	Patient Name	Patient MRN	Visit #	Age	Sex
Date of Birth	Attending Physician	Admit Date	Discharge Date		
Feb					

thiamine. He was found to be in acute kidney failure and received 3 liters of saline. He was started on bicarbonate drip, as he was found to have severe rhabdomyolysis with muddy brown urine output. He had been on Lipitor but it was unclear as to when this was last taken; he was unable to give a history. The Lipitor was stopped.

PAST MEDICAL HISTORY:

Hyperlipidemia, self-injurious behavior at age in to avoid serving in the military. He has multiple scars on his arms.

Table 22-4. Classification and differential diagnosis of renal failure

	Prerenal Azotemia	Postrenal Azotemia	Intrinsic Renal Disease		
			Acute Tubular Necrosis (Oliguric or Polyuric)	Acute Glomerulonephritis	Acute Interstitial Nephritis
Etiology	Poor renal perfusion	Obstruction of the urinary tract	Ischemia, nephrotoxins	Poststreptococcal; collagen-vascular disease	Allergic reaction; drug reaction
Urinary Indices					
Serum BUN:Cr ¹ ratio	>20:1	>20:1	<20:1	>20:1	<20:1
U _{NA} (mEq/L)	<20	Variable	>20	<20	Variable
FE _{NA} (%)	<1	Variable	>1	<1	<1; >1
Urine osmolality (mosm/kg)	>500	<400	250-300	Variable	Variable
Urinary sediment	Benign or hyaline casts	Normal or red cells, white cells, or crystals	Granular (muddy brown) casts, renal tubular casts	Dysmorphic red cells and red cell casts	White cells, white cell casts, with or without eosinophi

¹Blood urea nitrogen:creatinine ratio.

Proportions of Causes of Acute Renal Failure*

<u>Etiology</u>	Prerenal Azotemia	Postrenal Azotemia	Intrinsic Renal Disease (Acute Tubular Necrosis; Acute Glomerulonephritis; Acute Interstitial Nephritis)
<u>Proportion</u>	40 – 80%	5 – 10%	Up to 50% (ATN: 85%; AGN: 5%; AIN: 10 – 15%)

--Watnick and Morrison (2006)

Clinical Findings in Acute Renal Failure

“... The uremic milieu of acute renal failure can cause nonspecific symptoms, when present, they are often due to azotemia or its underlying cause... azotemia can cause nausea, vomiting, malaise, and altered sensorium... nonspecific acute abdominal pain and ileus as well as platelet dysfunction... neurologic examination reveals encephalopathic changes with asterixis and confusion; seizures may ensue...”

Watnick and Morrison (2006)

CASE PRESENTATION

- * A middle-age, retired male, former laborer, retired after winning a lottery award, lives with wife and mother-in-law
- * Longstanding history of idiopathic seizure disorder, intermittently treated and intermittently compliant/adherent, with a criminal history of two Domestic Violence (DV) episodes
- * Involved in DV episode over a general-intimate period during which he killed his wife and injured his mother-in-law with a knife
- * He claimed that his treating neurologist (not documented) had halved his dose of anticonvulsant about two months before the incident, and that he himself had no memory of the incident because it occurred during a seizure (“intraictal”)
- * A court-ordered psychiatric evaluation supported NGRI; a somatoforensic evaluation was arranged as a “second opinion”
- * A trial was held at which both mental health professionals testified

COMMON CHARACTERISTICS OF EPILEPTIC SEIZURES*

1. Epileptic seizures are usually discrete, time-limited events with an identifiable onset and termination
2. Most epileptic seizures, particularly those types which possibly could be implicated as a cause of ictal violence, have a well-defined and predictable evolution of behavior from beginning to end
3. After termination of most seizures, there is a progressive recovery of consciousness and neurological function
4. Epileptic seizures may be expressed as a variety of behaviors within one seizure type, but epileptic seizures are generally stereotyped within the same individual

**D. Tremain (2003)*

Ictal Aggressions: Pathophysiology*

- * Primary ictal aggression (“... directly stimulated by the epileptic discharge”)
- * Secondary ictal aggression (“... distribution of normal social controls by a seizure discharge...”)
- * Non-aggressive violent automatisms (“... a stereotyped automatism... not directed toward a person or object... no aggressive intent...”)
- * Resistive violence (“... reactive automatism or... a post-ictal confused state...”)
- * Post-ictal psychosis

**D. Tremain (1991)*

FORENSIC EVALUATION OF EPILEPTIC AGGRESSION*

1. What are the fundamental characteristics of epileptic seizures? How do we determine if a paroxysmal event is an epileptic seizure?
2. Under what circumstances could ictal aggression or violence occur? What is the pathophysiology of ictal aggression, if it occurs at all?
3. Is there evidence from the medical or legal literature that ictal aggression has actually occurred? What is that evidence?
4. Is there evidence that inter-ictal aggression occurs as a part of an epilepsy syndrome? Is epilepsy more frequent in violent prisoners than in the general population?
5. Are there other causes of paroxysmal violence which should be considered in a different diagnosis of ictal aggression?
6. What guidelines should be followed by an expert witness when considering the possible relationship between a violent event and an epileptic seizure?

**D. Tremain (2003)*

CRIMINAL RESPONSIBILITY

“Actus non facit reum nisi mens sit rea”

(“The deed does not make a man guilty unless his mind is guilty”)

--Quoted in D. Tremain (2003)

CLINICAL AND FORENSIC ANALYSIS CONTRASTED

For the clinical practitioner, the conceptual framework is some variation of identification, chief complaint, history of the present illness, pertinent past history, laboratory test data, differential diagnosis, medical diagnostic impression, and treatment plan.

For the forensic psychiatric practitioner, the four-step conceptual framework is issue, legal criteria, relevant data, and reasoning process.

CLINICAL FORENSIC ANALYSIS*

For the clinical practitioner, the conceptual framework is some variation of identification, chief complaint, history of the present illness, pertinent past history, laboratory test data, differential diagnosis, medical diagnostic impression. For the forensic psychiatric practitioner, the four-step conceptual framework is issue, legal criteria, relevant data, and reasoning process:

1. *Issue*: What is the specific psychiatric-legal issue to be considered?
2. *Legal criteria*: In the jurisdiction in which this specific psychiatric-legal issue must be resolved, what are the legally defined terms and criteria that will be used for its resolution?
3. *Relevant data*: Exactly what information (such as part of what might be collected by a clinician following the traditional clinical framework for data organization) is there that is specifically pertinent to the legal criteria that will be used to resolve the specific psychiatric-legal issue?
4. *Reasoning process*: How can the available relevant data be applied to the legal criteria so as to yield a rationally convincing psychiatric-legal opinion?

*R. Rosner (2003)

BASEBALL, UMPIRES AND THE LAW

In baseball, there are three types of umpires, which correspond to three roles in the legal system, as follows:*

TYPES OF UMPIRES

“I call it as I see it.”

“I call it as it is.”

“It’s nothing until I call it.”

ROLES IN THE LEGAL SYSTEM

The lawyer (advocate)

The expert witness

The judge (court)

*Anon., ca 2008

THE CORE CLINICAL QUESTION FOR THESE CASES

Were the individuals actively psychotic at the times of the offenses, or were they angry and exercising bad judgment in connection with the offense(s)?

THE VOTE

- * Guilty (or responsible) as charged
- * Not guilty
- * Not guilty by reason of insanity (“NGRI”)
- * Need more information
- * Don’t know

THE ANSWERS

- * Neurosyphilis: NGRI (bench trial)
- * AIP: Child was returned to her
- * Huntington's Disease: Diagnosis was forensically irrelevant
- * ARF: pending
- * Epilepsy: Guilty (jury trial)



THANK
YOU