Neurocritical Care Society

ENLS®

EMERGENCY NEUROLOGICAL LIFE SUPPORT

What To Do in the Critical First Hours of a Neurological Emergency

VERSION 5.0

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The ENLS Course presents a stepwise approach to clinical care in the “golden hour(s)” of a neurocritical care emergency. The 14 ENLS topics span the broad range of neurologic emergencies and cover aspects of general emergency medicine and critical care that need to be specifically tailored to the patient with acute nervous system illness or injury. Each module contains an initial algorithm, a checklist of important clinical points, a list of information needed for communication to improve transitions across care settings, clinical pearls, and more. ENLS is relevant for a wide array of care providers, from prehospital to specialist. Content is research-based and is updated every two years.

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Emergency Neurological Life Support: Approach to the Patient with Coma

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Abstract

Coma is an acute failure of neuronal systems governing arousal and awareness and represents a medical emergency. When encountering a comatose patient, the clinician must have an organized approach to detect remediable causes, prevent neurologic injury, and determine a hierarchical approach for diagnostic testing, treatments, and neuromonitoring. Coma was chosen as an Emergency Neurological Life Support (ENLS) protocol because timely medical and surgical interventions can be lifesaving and need implementation in a rapid manner. The initial work-up of such patients is critical to establishing a correct diagnosis.

Key words: Coma, Consciousness, Critical Care, Neurocritical Care, Encephalopathy

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1 Introduction

Coma is characterized by the absence of arousal (wakefulness, vigilance) and awareness of self and environment, lasting for more than one hour.[1] Certain causes of coma are readily identified, while others may require extensive testing to discover an etiology and it is important that diagnostic and therapeutic steps occur simultaneously.[2]

The ENLS suggested algorithm for the initial management of coma is shown in Figure 1. The initial step is to assess for coma and reversible conditions while incorporating available history and physical exam findings to best diagnose and concurrently treat patients with acute coma. Table 1 has a list of suggested items to complete within the first hour of patient evaluation; this is meant to provide a framework for the principles of diagnosis and emergent management of coma, which can be adapted to reflect global and regional variations based on the local availability of diagnostic tools and treatments.

TABLE 1
Coma checklist for the first hour

Checklist
☐ Evaluate/treat circulation, airway, breathing and cervical spine
☐ Exclude/treat hypoglycemia or opioid/benzodiazepine13 overdose
☐ Serum chemistries, arterial blood gas, urine toxicology screen
☐ Emergent cranial CT (and CT Angio Brain if appropriate) to determine if coma etiology is structural or vascular in etiology

2 Prehospital Considerations

If coma is identified in the prehospital setting, initial evaluation and management steps as outlined in this protocol should be implemented as soon as possible to maximize chances of neurologic recovery. Emergency medical services (EMS) teams will assess airway, breathing, and circulation (ABCs); check Glasgow Coma Scale[3] (GCS), pupils, and vital signs, including blood glucose, and obtain intravenous (IV) or intraosseous (IO) access. Based on assessment findings, EMS teams will establish an airway, ventilate if needed, check for and correct hypoglycemia, administer naloxone, and treat for shock with fluids and pressors if indicated. In addition, prehospital personnel should collect pertinent information from witnesses and from contextual or environmental observations. Witnesses may be able to provide information on the time course of the neurological decline as well as any prodromal symptoms. Family or friends may have information regarding the patient's past medical history, prescribed medications, drug or alcohol use, any history of recent illness that might suggest an infectious cause, signs that might suggest a drug or medication overdose, seizure activity, or recent trauma. A quick look around the site in which the patient was found can yield valuable information including signs of trauma or environmental exposures, current prescription bottles, empty pill bottles, drugs, or alcohol that might suggest overdose or intoxication.

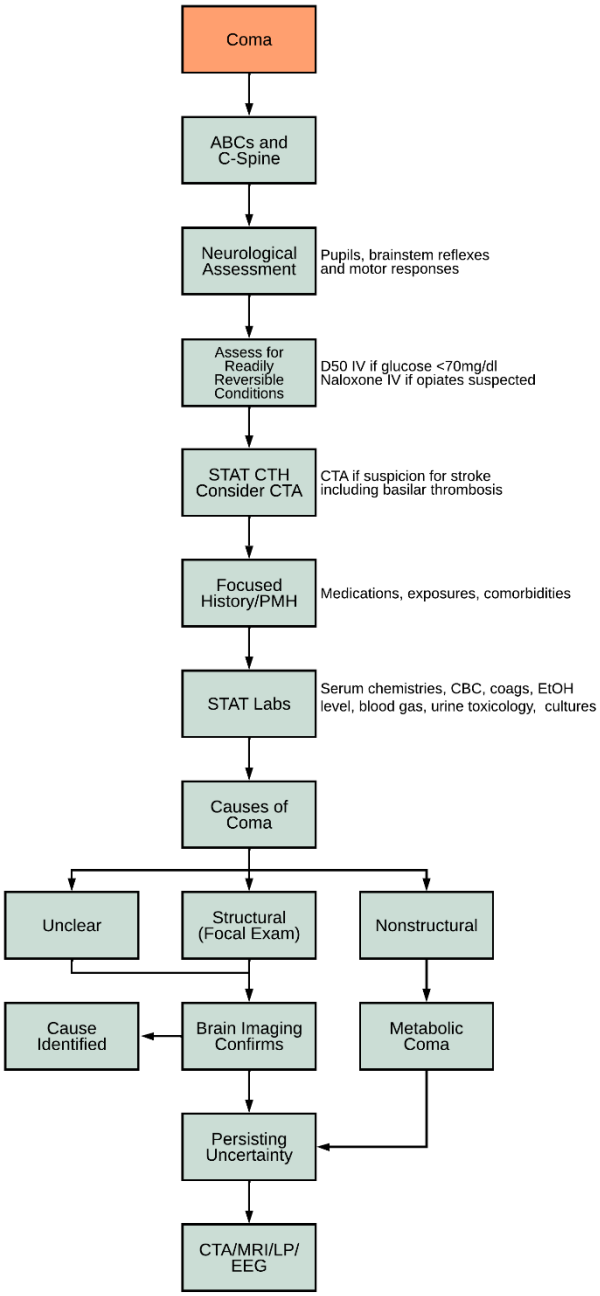


FIGURE 1: ENLS Approach to the Patient with Comaprotocol

TABLE 2
Prehospital checklist and handoff

Prehospital checklist and handoff
• Airway, breathing, ventilation issues
• GCS, pupils, and vital signs on presentation
• IV or IO access, site and patency
• Ruled out Hypoglycemia
• History from bystanders, witnesses, contextual or environmental observations (pill bottles, signs of trauma or seizure activity)
• Naloxone, dose administered and response
• Medications administered, dose and response (naloxone, benzodiazepine, dextrose etc.)
• Time when patient was last seen normal
• Prodromal symptoms when last seen

2.1 ABCs and Cervical Spine Precautions

The unconscious patient's ABCs should be quickly assessed and concurrently treated (see the *ENLS Airway, Ventilation, and Sedation* protocol). Verifying patency of the airway is an overriding initial priority to ensure adequate oxygenation and ventilation. The patient's cervical spine should be immobilized if the possibility of injury cannot be ruled out. A rapid initial survey should follow to evaluate for notable physical findings of the head, neck, chest, abdomen, and extremities.

IV or IO access should be established during the initial evaluation and, if possible, in the prehospital environment. Prompt blood glucose testing should be performed in all unconscious patients. Table 3 lists pharmacological treatment that are administered for common toxic/metabolic causes of coma, as well as elevated ICP. These should be administered in the prehospital setting based on syndromic presentation. Table 9 lists common clinical findings for these toxidromes; elevated ICP often requires a high index of suspicion but may also involve pupillary changes or the Cushing's triad of (1) bradycardia, (2) hypertension with widened pulse pressure owing to increased systolic pressure and decreased diastolic pressure, and (3) irregular respiration.[3]

3 General And Neurologic Examination

A general physical examination should be performed including assessment of vital signs/respiratory patterns, facial and motor symmetry, and cranial and peripheral reflexes.[4] If hypotension is present, the cause should be pursued while fluid repletion and/or vasopressors are started. Blood pressure elevation in the comatose patient may be a sign of an underlying life-threatening process, such as elevated intracranial pressure (ICP) or acute stroke, which must be identified and treated promptly. Extremely high blood pressure should be treated if intracranial hemorrhage is suspected as a cause of coma (history of anticoagulation, fall).[5] A search for signs of trauma and other conditions that might require emergent surgical or medical management are central goals of the initial survey.

TABLE 3
Prehospital pharmacological therapy for coma

Cause of coma	Pharmacological therapy
Hypoglycemia	Dextrose 50% 20-50ml IV (peds) 25% 2-4ml/kg 10% Dextrose 50-100ml IV (peds: 5-10ml/kg) can be given if D50 unavailable Thiamine should be given prior to dextrose in patients with risk factor for nutritional deficiency (e.g., chronic EtOH use, bariatric surgery, malabsorption states) or if history unknown
Opioid overdose	Naloxone 0.04-0.4 mg IV/IM or 1-2 mg per nare into both nares, can be repeated every 2-3mins for desired degree of counteraction. If initial use intranasal, switch to IV/IM when possible.
Anticholinergic toxicity	Physostigmine 0.5-2 mg slow IV push with rate not to exceed 1 mg/min. Dose can be repeated in 30-60mins if clinically effective to ameliorate symptoms. Atropine 1-2 mg IV must be kept ready nearby for immediate use if bradycardia or other signs and symptoms of cholinergic excess. Dose can be repeated every 3-5mins if previous dose did not induce a response
Elevated ICP	Hypertonic saline 3% 5ml/kg over 5-20min (range 2.5-5ml/kg) can be given through peripheral IV as 250ml bolus over 30mins or 500ml bolus over 60mins.
	Mannitol 20-25% dose 0.5-1 gm/kg over 5-15min, can be redosed every 4-6 h. Caution must be exercised in heart and renal failure due to the high osmotic load infused and lower doses (0.25-0.5 g/kg) may be used. Requires in-line filter (precipitates-crystal formation); may require warming to dissolve crystals before administration.

The emergency neurological assessment of the unconscious patient has four parts[1]: level of consciousness (LOC), brainstem assessment, evaluation of motor responses, and appraisal of breathing patterns. Arousal is assessed by looking for spontaneous opening of the eyes, visual fixation or pursuit (tracking), and spontaneous and purposeful movements of the extremities. Altered motor strength can also be tested via resistance to movement. The examination of a comatose patient should involve increasing intensity of stimulation until a response is evoked or it is deemed that the patient is unable to respond. Start with a simple verbal cue (e.g., "Are you ok?") and progress to louder voice, physical stimuli, and noxious stimuli. Noxious stimuli could include a sternal rub, nail bed pressure or trapezius muscle squeeze.

The LOC can be expressed quantitatively by the GCS in adults and children (see Tables 4 and 5).[3] The GCS is most valuable for trending sequential LOC examination responses of a particular patient. However, the GCS is limited by its inability to account for alterations in brainstem function, hemiparesis, or aphasia or help differentiate between different etiologies of coma. Patients with identical total GCS scores may have very different clinical presentations due to different combinations in the motor, verbal, and eye sub-scores. The Full Outline of UnResponsiveness (FOUR) score incorporatesmore

detailed information on brainstem responses and has been validated in a variety of clinical settings (Figure 2).[5–7] Use of the FOUR score tool can assist the clinician in determining the presence of a locked-in state vs. a true vegetative state.[8] Similar to the GCS, each section is scored separately and allows for a trending of LOC.

TABLE 4
Glasgow Coma Scale (GCS) for use in adult patients[1]

Glasgow Coma Scale (GCS)
Eye opening
Spontaneous - 4
To speech – 3
To pain - 2
No response - 1
Best motor response
Obeys - 6
Localizes – 5
Withdraws – 4
Abnormal flexion- 3
Abnormal extension - 2
No response - 1
Best verbal response
Oriented - 5
Confused conversation - 4
Inappropriate words- 3
Incomprehensible sounds - 2
No response - 1

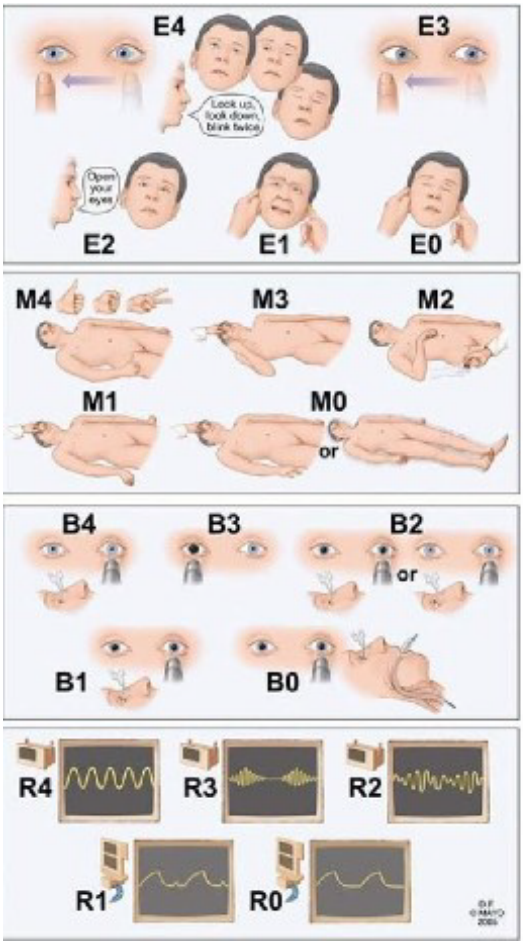
*GCS range 3–15

3.1 Cranial Nerve Testing

Testing of the cranial nerves is focused on assessing the integrity of the afferent limb of the brainstem reflex being tested, brainstem nuclei involved and the efferent tracts.[9] Brainstem reflexes are key to the initial coma evaluation and include pupillary assessment (size, reactivity, and symmetry), corneal reflex, response to visual threat, oculocephalic reflex (performed only if cervical trauma or instability is not a consideration), gag and cough reflex. Pinpoint pupils are suggestive of pontine damage, usually from hemorrhage or ischemic infarction. Enlarged and nonreactive pupils suggest damage to the midbrain or compression of the third cranial nerve. Pupillary changes can also be suggestive of a drug overdose (see Table 6). Spontaneous roving eye movements suggest bilateral cortical dysfunction and an intact brainstem. Dysconjugate resting gaze may occur with structural or metabolic processes or extraocular movement impairment from cranial 3^{rd}, $4^{th,}$ or 6^{th} nerve palsies. Psychoactive and anti-seizure drugs have been associated with depressed vestibulo-ocular responses. Jerky, nystagmoid movements may indicate non-convulsive status epilepticus or brainstem or cerebellar ischemia. Fundoscopy may reveal retinal hemorrhages or papilledema if there has been prolonged increased ICP; more acutely, high ICP may manifest as optic disc blurring or lack of spontaneous venous pulsations in

TABLE 5
Pediatric Glasgow Coma Scale (PGCS)[1] Adapted with permission Pediatric Glasgow Coma Scale PGCS

Pediatric Glasgow Coma Scale PGCS				
		>1 year	<1 year	Score
Eye opening		Spontaneously	Spontaneously	4
		To verbal command	To shout	3
		To pain	To pain	2
		No response	No response	1
Motor response		Obeys	Spontaneous	6
		Localizes pain	Withdraw to touch	5
		Flexion-Withdrawl	Flexion-Withdrawl	4
		Flexion-abnormal (decorticate rigidity)	Flexion-abnormal (decorticate rigidity)	3
		Extension (decerebrate rigidity)	Extension (decerebrate rigidity)	2
		No response	No response	1
	>5 years	2-5 years	0-23 months	
Verbal Response	Oriented	Appropriate words/phrases	Smiles/coos appropriately	5
	Disoriented /confused	Inappropriate words	Cries and is consolable	4
	Inappropriate words	Persistent cries and screams	Persistent inappropriate crying and/or screaming	3
	Incomprehensible sounds	Grunts	Grunts, agitated and restless	2
	No response	No response	No response	1
Total Pediatric Glasgow Coma Score (3-15)				



EYE RESPONSE

4 = Eyelids open or opened, tracking or blinking to command
3 = Eyelids open but not to tracking
2 = Eyelids closed but opens to loud voice
1 = Eyelids closed but opens to pain
0 = Eyelids remain closed with pain stimuli

MOTOR RESPONSE

4 = Thumbs up, fist, or peace sign
3 = Localizing to pain
2 = Flexion response to pain
1 = Extension response
0 = No response to pain or generalized Myoclonus status

BRAINSTEM REFLEXES

4 = Pupil and corneal reflexes present
3 = One pupil wide and fixed
2 = Pupil or corneal reflexes absent
1 = Pupil and corneal reflexes absent
0 = Absent pupil, corneal, or cough reflex

RESPIRATION

4 = Regular breathing pattern
3 = Cheyne-Stokes breathing pattern
2 = Irregular breathing
1 = Triggers ventilator or breathes above ventilator rate
0 = Apnea or breathes at ventilator rate

FIGURE 2: Four Score

the optic disc. A thorough brainstem exam can uncover early signs of basilar stroke and allow early therapies capable of minimizing long term disability. Brain stem reflexes may be absent in hypothermic patients, up to a few hours after cardiac arrest, or in patients who have received neuromuscular paralysis.

Motor function is assessed by observing spontaneous movements, responses to verbal command or noxious stimulation or for posturing. Symmetric posturing, either extensor ("decerebrate") or flexor ("decorticate"), may occur in either structural or metabolic coma. Generalized or symmetric findings raise the possibility of a toxic or metabolic process that involves brainstem or thalamic and brainstem arousal centers. Muscle tone of the extremities may be assessed by passive movement of the limbs, mainly elbow and knee flexion. The examiner should distinguish purposeful movements from reflex activity. Examples of purposeful activity include following commands: axial (sticking out tongue) or acral (showing two fingers or thumbs up), pushing the examiner away, reaching for the endotracheal tube, or localizing to noxious stimulation. Examples of reflexive activ-

TABLE 6
Pupillary changes reflecting underlying etiology[1]

Pupillary change	Possible etiologies/localization
Pinpoint pupil	Opioids Cholinergic intoxication Pontine damage (interrupts descending sympathetic pathways)
Dilated, non-reactive pupils	Cerebral anoxia, global Barbiturates Atropine Hypothermia Brain death
Dilated, reactive pupils	Pretectal lesions Stimulants (cocaine, methamphetamine), hallucinogens including PCP/LSD
Anisocoria (pupillary asymmetry)	3rd nerve compression from uncal herniation Localized drug effect (e.g., ipratropium, tropicamide)
Mid-position, fixed or irregular	Midbrain lesion

ity include withdrawal, flexion or extensor posturing to noxious stimulation. Ability to grasp should not be considered following commands without the ability to reproducibly let go on command, as this too can be a reflex rather than a conscious movement. Deep tendon reflexes should be performed with particular attention to briskness and symmetry of findings.

Observed breathing patterns may also have localizing value in the evaluation of coma. Lesions of the pons or midbrain can result in neurogenic hyperventilation. Central hyperventilation may also be a sign of underlying acidosis and compensatory respiratory alkalosis. Cluster (Biot's) breathing may be seen resulting from a pontine lesion. Ataxic or absence of spontaneous breathing (apnea) may be seen in medullary injury (Chart 1).[1]

4 Focused Presenting and Past Medical History

Historical information elicited from witnesses, friends, family, co-workers, or EMS personnel may suggest the cause of coma. EMS personnel may have valuable details about the circumstances in which the patient was found. The time course of the alteration of consciousness may be helpful in suggesting an etiology. An abrupt or acute onset of symptoms suggests a stroke, seizure, or a cardiac event with impaired cerebral perfusion. A more subacute onset of encephalopathy evolving into coma could suggest a metabolic or possibly infectious process. Past medical, surgical, or psychiatric history; alcohol or illicit drug use history; and any environmental toxic exposures should be included in the information gathering. Medication history is paramount in identifying a possibility of overdose and may also provide valuable clues to the medical history in the absence of

Respiratory patter	Pattern	Localization
Cheyne-Stokes		Global/metabolic encephalopathy Impaired forebrain or diencephalon
Central neurogenic hyperventilation		Metabolic encephalopathy High brainstem tumors (rare)
Apneusis		Bilateral pontine lesions
Cluster breathing/ataxic breathing		Pontomedullary junction lesions
Apnea		Lesions affecting ventrolateral medulla bilaterally (ventral respiratory group)

CHART 1: Respiratory patterns reflecting underlying etiology [1]

other sources.[10] The electronic medical record may provide rapid access to the patient's past medical history if the patient can be reliably identified.[11]

5 Recommended Labs

Unless a readily reversible cause of unresponsiveness, such as hypoglycemia, has been discovered and corrected, additional laboratory testing should be obtained. Serum chemistries, a basic hematological panel, and blood gas analysis should be considered. Point of care (POC) testing should be utilized where available. Co-oximetry may be beneficial in selected patients suspected of carbon monoxide poisoning. Toxicology testing such as ethanol level and urine toxicology screen should be obtained, though variability and availability of exhaustive toxicology screens is limited in emergent settings. Microbiologic studies, including cultures of blood and urine, are helpful in many cases.

6 Initial Formulation: Structural, Non-Structural, or Unclear Causes of Coma

Information obtained during initial stabilization, physical assessment, neurological assessment, focused history, and stat laboratory results will typically help in classifying patients into likely structural or non-structural causes of coma and direct further investi-