Pedro Orta, MPH, and Brad Cole, MD

GENERAL TIPS

- Examine the newborn in its caregiver's lap or on the exam bed.
- Take advantage of every opportunity to observe the child at rest. Assess for symmetrical movement of all extremities and its interactions with the environment (may be minimal the closer to birth).
- Start with steps that require the infant to be quietly resting, not crying. This is particularly important while examining the fontanels, tone and reflexes.
- Reserve potentially irritating steps for the end. These include measuring head circumference, fundoscopy, corneal and gag reflexes and sensory testing.

I. History

- A. A detailed history is often the most important part of the neurological exam.
- B. Assess the 7 Dimensions of the Chief Complaint: Chronology, Location, Quantity, Quality, Aggravating / Alleviating Factors, Setting, and Associated Manifestations. A good acronym is "<u>Cute Ladies Quilting Quilts Black and White So Amazingly</u>."
- C. Get a good **Birth History** including gestational age at birth, complications during pregnancy (including infections), maternal substance use, Apgar scores, complications during delivery, NICU admission, post-birth complications, etc.
- D. Review the patient's **Past Medical History** including immunization status, accidents, allergies, and other medical conditions. Particularly focus on <u>past</u> <u>neurological events</u>: head trauma, seizures, status epilepticus, meningitis, etc.
- E. Review the **Medications** that both the patient and their mother (if breastfeeding) are currently taking. If discussing seizures, ask about <u>adverse effects</u>.
- F. Assess **Developmental Milestones** using the Denver II Assessment Tool.
- G. Review any Family History of neurological disorders.

II. General Physical Examination of the Newborn (focused on Neurological Connections)

A. Your physical exam should start by observing the newborn un-obtrusively while it is resting in its caretaker's arms or in its bassinette. Much can be learned from the child's **general appearance**. Skilled observation of morphological features, mental

status, behavior, awareness of and interaction with environment, posture, and movement can give clues of neurological pathology.

- B. A detailed **examination of the scalp** is essential to the neuro exam.
 - 1. Visualize the skull for abnormalities indicative of neuropathology:
 - a. Occipital prominence \rightarrow Dandy-Walker syndrome.
 - b. Scalp vein prominence \rightarrow increased intracranial pressure (ICP).
 - c. Asymmetry or abnormal placement of the eyes and ears may be phenotypic of various neurodevelopmental disorders.

TABLE 1: 3 & 9 Rule		
Age	Circum.	
Newborn	35 cm	
3 months	40 cm	
9 months	45 cm	
3 years	50 cm	
9 years	55 cm	

- 2. **Measure** <u>head circumference</u> at the point of greatest occipitofrontal circumference and is plotted on a standard growth curve.
 - a. Deceleration in growth relative to the standard curve over time may indicate anoxic brain injury or degenerative neurologic disorder; acceleration in growth may indicate hydrocephaly.
 - b. Macrocephaly and microcephaly are respectively defined as a head circumference greater and lesser than 2 SD from the mean.
 - c. An estimate for head circumference is the 3 & 9 rule (Table 1).
- 3. **Palpate** the skull with particular attention to the suture lines. The **posterior** and **anterior fontanels** are usually patent until 3 months and 2 years of age, respectively. These should be slightly depressed, soft and pulsatile when an infant is calm. The fontanels should be examined when the infant is calm to avoid possibly masking actual pathology.
 - A bulging, tense fontanel may indicate increased ICP and may be associated with infections, tumors and other space occupying lesions. The fontanel may also bulge during crying or fevers.
 - b. Premature suture closure may occur in craniosynostosis.
 - c. Suture separation (cracked pot sign) may indicate increased ICP.
- 4. **Percuss** the entire surface of the skull. Tenderness to percussion may indicate osteomyelitis.
- 5. Auscultate the skull for bruits in six locations: the bilateral globes, temporal fossae, and retro-auricular / mastoid areas. Intracranial bruits can be heard in angiomas, anemia, thyrotoxicosis and meningitis.
- 6. The **ocular exam** is an essential element of the neonatal neuro exam and must include evaluation for epicanthal folds, coloboma, conjunctival telangiectasias and cataracts. Ophthalmoscopic evaluation of the optic disk may reveal papilledema (associated with increased ICP), retinal hemorrhages (suggestive of non-accidental trauma) or cherry-red spots.

- 7. Finally, assess the **cranial nerves** as described below.
- C. Now proceed with **general physical examination** of the <u>undressed</u> newborn. Special care should be taken while examining the skin, neck, spine, hands and feet (described below). Other sections of the physical exam are not discussed.
 - 1. The **skin** and nervous system both originate from the ectoderm. As such, many neurological pathologies have cutaneous manifestations.
 - a. Common neurocutaneous lesions can be seen in Fig. 1-3.
 - b. Hair and nail quality may also indicate neuropathology. E.g. the friable, kinky hair characteristic of Menkes kinky hair disease is also associated with mental retardation and optic atrophy.
 - The newborn's neck and spine should be examined for midline defects myelomeningocele, cutaneous dimples, sinus tracts, subcutaneous lipomas, hair tufts – that may suggest neural canal abnormalities.
 - 3. Assess both **hands and feet** for signs of neurodevelopmental pathology. Examples include the single palmar creases and increased spacing of the big toes associated with downs syndrome (Fig. 4-5).
 - 4. Note **unusual odors** that may indicate inborn errors of metabolism.

III. The Focused Neuro Exam

- A. The purpose of the neuro exam is to localize the region of the nervous system from which the newborn's neurological symptoms arise. The links between neuro exam segment and nervous system localization can be conceptualized as follows:
 - Mental Status Exam ightarrow cerebral cortex function
 - Cranial Nerve Exam \rightarrow brainstem function
 - Motor Exam \rightarrow upper and lower motor neuron function
 - Sensory Exam \rightarrow peripheral sensory receptors and central connections
 - Deep Tendon Reflexes \rightarrow UMN inhibition of tendon stretch reflexes
 - Gait / Extremity Movement \rightarrow functional assessment of motor system

B. Mental Status

1. The <u>normal mental status</u> of the neonate is characterized by quiet, intermittent wakefulness and occasional irritability (usually due to hunger, pain or discomfort) interspersed with extended periods of sleep. Persistent irritability or lethargy indicate that something is wrong.

- 2. Newborns have minimal meaningful <u>interaction with their environments</u> but do retract to painful stimuli, blink in response to sudden loud noises, and progressively focus their eyes on objects within their field of vision.
- 3. <u>Language function</u> develops with maturity. Usually, infants start quietly cooing around month three. They start to recognize familiar voices and respond by smiling around this time. Other indications of expressive and responsive language function do not develop until after six months, with the ability to say simple two syllable words like 'mama' and 'papa' initiating around one year of age. Until then, it is difficult to diagnose <u>expressive (Broca) and receptive (Wernicke) aphasias</u> that respectively present as spares, non-fluent language and fluent, nonsensical speech.

C. Cranial Nerve Exam

- 1. **CN I** is responsible for <u>the sense of smell</u>. Smell begins to develop at 5 to 7 months of age but is difficult to assess in non-verbal children younger than 2 years. In children older than 2, smell should be assessed with aromatic substances instead of volatile substances (like ammonia) that could irritate the nasal and oral mucosa.
- 2. **CN II** is the optic nerve. While <u>visual acuity</u> in newborns is estimated to be 20/200 and improve to 20/20 by six months of age, it is also difficult to assess visual acuity with standard charts in non-verbal children.
 - a. <u>Peripheral vision</u> is tested by observing the reaction when an object is covertly brought into the field of the vision from behind the infant's head. Full-term newborns are able to follow objects.
 - b. <u>Anterior tract lesions</u> affecting the retina, optic nerve, and optic chiasm are diagnosed with the *swinging flashlight test*. When light is pointed at the affected eye both eyes inappropriately dilate, but appropriate bilateral constriction occurs when light is pointed at the normal eye. This is known as an *afferent pupillary defect* or *Marcus Gunn pupil*. <u>Posterior tract defects</u> result in loss of visual fields that are difficult to assess in nonverbal children.
 - c. <u>Lesions in the sympathetic innervation of the pupil</u> result in *Horner's Syndrome* (ptosis, miosis and ipsilateral facial anhidrosis).
- 3. **CN III, IV, & VI** are assessed through careful observation of the neonate's voluntary eye movements, particularly focusing on preferential gaze.
 - a. CN III (the oculomotor nerve) is responsible for moving the eye up and in. In CN III lesions, the involved eye will be positioned down and out. There will also be associated ptosis and pupillary dilation.
 - b. CN IV (the trochlear nerve) moves the eye upward. Injury to CN IV results in downward deviation of the affected eye.

- c. CN VI (the abducens nerve) moves the eye outward. Injury results in medial deviation of the affected eye. Due to its long intracranial conduit, CN VI palsy is a nonspecific indication of increased ICP.
- 4. **CN V** mediates facial sensation. It can be assessed in neonates by gently brushing the cheek with your finger. If functional, the *rooting reflex* (explained below) will be elicited. The ophthalmic division of CN V and VII can be tested with the corneal reflex.
- 5. **CN VII** can be assessed by observing the newborn's face during crying, resting and blinking. A peripheral lesion causes weakness of the ipsilateral facial muscles. A central lesion causes contralateral weakness.
- 6. CN VIII lesions result in vertigo, deafness, and tinnitus.
 - a. Injury to the **vestibular** portion of the nerve results in vertigo, vomiting and nystagmus that may manifest as irritability in the non-verbal newborn. Nystagmus can be assessed by observing involuntary beating of the eye, usually with a fast phase in one direction and a slow phase in the other.
 - b. Auditory function can be assessed in newborns by observing blinking in response to an abrupt, loud sound. By two months, infants should smile in response to familiar voices. By four months, infants will turn their heads in the direction of the sound.
- 7. CN IX & CN X lesions result in absence of the gag reflex (normally present at all ages, except in extreme prematurity). Weak sucking, drooling, gagging and nasal regurgitation may indicate CN X dysfunction.
- 8. **CN XI** can be assessed by observing the posture of the head, neck and shoulders. Persistent drooping of the shoulder or tilting of the head may indicate dysfunction of CN XI, the trapezius or sternocleidomastoid.
- 9. **CN XII** dysfunction causes deviation of the tongue towards the lesion. Atrophy and fasciculation of the tongue, indicating injury at the anterior horn cell as in spinal muscular atrophy, are best observed during sleep.
- D. **Motor Nerve** function is assessed by passive observation of spontaneous movement of the extremities as newborns are unable to follow commands.
 - 1. **Power** is assessed by observing spontaneous, symmetrical movement of extremities. Best done by holding the infant in a supine position with support at the buttocks and shoulders. Power is graded on a 0-5 scale:
 - 5. Normal movement against resistance and gravity
 - 4. Weak movement against resistance

- 3. No movement against resistance but normal movement against gravity
- 2. No movement against gravity
- 1. Minimal movement without gravity or resistance
- 0. Complete paralysis
- 2. **Tone** refers to the dynamic resistance of muscles to passive extension. It should be assessed while the infant is at rest and not crying.
 - a. Lower motor and cerebellar conditions are characterized by decreased tone (hypotonia). While upper motor neuron (cortex and pyramidal tract) lesions result in increased tone (hypertonia).
 - Hypotonia can be assessed using <u>the scarf test</u>. Pull the infant's arms across their chest. If the elbow crosses the midline, hypotonia is present.
 - 2) Hypertonia and rigidity are well exemplified by spastic cerebral palsy, which affects either the motor cortex or the pyramidal tracts that relay UMN signals to muscles.
 - b. See Horizontal and Vertical Suspension Reflexes in Table 2.
- 3. **Posture** is assessed in the supine position and normally progresses with decreased flexion of the extremities (first the lower, then the upper extremities) and increased tone of the trunk and neck over time.
 - a. Before 32 weeks of gestation, premature newborns present with full extension of all extremities. At 32 weeks of gestation, the lower extremities start to flex through term when full lower extremity (hips, knees) flexion and upper extremity extension is the norm.
 - b. Flexion starts to decrease at 3 months.
 - c. **Recoil**, the tendency of extremities to return to normal position when flexed or extended, is absent in extreme prematurity.
- 4. **Bulk** refers to the volume of muscle tissue. Bulk may be diminished in many LMN diseases (e.g. neuropathies) and increased in other conditions (e.g. myotonia congenita, a genetic disease characterized by the inability of muscles to quickly relax after voluntary movements).
- 5. **Coordination** and ataxia are difficult to assess in newborns as they are incapable of intentional movement.
- 6. **Gait** cannot be assessed in neonates as they do not crawl or walk. Instead, one can assess symmetrical movement of all extremities. Subtle differences in power, tone, or coordination may indicate motor deficits.

- E. **Sensory Functions** are tested by assessing response to tactile, visual, and auditory stimuli. Aside from blinking with loud noises or retraction to painful stimuli, it is difficult to assess sensation in the newborn.
- F. **Deep Tendon Reflexes** are defined as involuntary extensions of the extremities in response to muscle sheath or tendon stretch. DTRs are increased in UMN lesions (due to decreased reflex inhibition) and decreased in LMN lesions. In addition to neurological factors, DTRs can be influenced by metabolic factors and anxiety.

Reflex	Spinal Nerve Roots	
Biceps	C5, C6	
Brachioradialis	C6	
Triceps	C7	
Patellar	L4	
Achilles Tendon	S1	

1. The main spinal nerve roots tested in DTRs are

- 2. DTRs are reported on a 5-point scale:
 - 0: Absent
 - 1+: Trace
 - 2+: Normal
 - 3+: Brisk
 - 4+: Non-sustained Clonus (repetitive vibratory contraction of the muscle in response to muscle and tendon stretch)
 - 5+: Sustained Clonus
- G. Primitive Reflexes (Table 2) are present at the time of birth and decrease over time. They represent spinal reflexes that decrease as the cerebral cortex develops its inhibitory functions and suppresses them. Asymmetry, persistence or recurrence of these reflexes may indicate a focal brain or peripheral brain lesion. For example, the grasp and rooting reflexes are inhibited as the frontal cortex matures and may reappear with focal lesions of the frontal cortex later in life.

Deflect	Deseriation	Age at	Age at
Reflex	Description	Appearance	Disappearance
Sucking Reflex	Touching the roof of the newborn's mouth with a	Birth	3 mo
	finger, pacifier, or nipple causes the newborn to		
	begin sucking. Decreased or absent in premature		
	infants prior to 36 weeks of gestation.		
Trunk Incurvation	Stroking the side of the infant's vertebrae results in	Birth	4 mo
(Gallant)	a deviation of the spine in the direction of the touch		
Rooting	Touching corner of infant's mouth results in mouth	Birth	4 – 6 mo
	(head) mildly turning in the direction of stimulus.		
Placing	Stroking the dorsum of the infant's foot with a	Birth	4 – 6 mo
(Stepping)	surface results in the infant attempting to place		
	their foot on that surface.		
Moro	Slight drop of head results in sudden extension	Birth	6 mo
	followed by flexion of upper and lower extremities.		
Grasping	Placing finger in the palm of an infant's hand results	Birth	6 mo
	in flexing of infant's fingers.		
Tonic Neck	While the infant is lying flat, turning of the head	Birth	6 mo
(Fencing)	causes extension of the ipsilateral extremities (left		
	extremities extend with leftward turn of head) and		
	flexion of the contralateral extremities (right		
	extremities flex with leftward turn of head).		
	Asymmetrical, prolonged responses are abnormal.		
Babinski	Scratching the plantar surface of the infant's foot	Birth	12 mo
	causes dorsiflexion of the great toe and fanning of		
	the toes. A <u>symmetric</u> response can be normal		
	through the age of 1 year.		
Oculocephalic	While the infant's eyes are open, turn its head left	Birth	
vestibular reflex	or right. If the brainstem is intact, their eyes should		
	move in the opposite direction of head movement.		
Vertical	Holding the infant at the chest in the upright	Birth	
suspension	position with both hands, gently elevate the infant		
	off the bed. Hyperextension (scissoring) of the legs		
	indicates hypertonia (spasticity). Consider CP.		
Horizontal	With the infant in the ventral position, hold the	Birth	
(ventral)	infant at the trunk and lift the infant. Normally, the		
suspension	spine will extend, and their eyes will look forward		
(Landau reflex)	just below the horizontal. Hypotonia is present if		
	the spine collapses in an upside-down "U" shape.		
Parachute	Moving the infant down towards a table while the	8 – 10 mo	
	infant is held at the chest facing down results in		
	extension of the arms as if to protect itself.		

Figures:



Figure 1: café au lait macules (flat light brown macules) associated with neurofibromatosis (NF type 1)



Figure 2: adenoma sebaceum (fibrovascular lesions that look like acne) associated with tuberous sclerosis



Figure 3: Shagreen patches (flesh-colored soft plaques with prominent follicular openings) associated with tuberous sclerosis



Figure 4: Simian (single) palmar crease associated with down syndrome



Figure 5: Sandal gap abnormality associated with down syndrome

Reference:

Marcdante, Karen J. and Robert M. Kliegman. "Neurology Assessment." *Nelson Essentials of Pediatrics*. 7th Ed. Philadelphia, PA: Elsevier Inc, 2015. Pages 612 – 616. Print.