
The Neurologic Exam in Neonates and Toddlers

2

Alison S. May and Sotirios T. Keros

Introduction

Performing a comprehensive neurological examination in children is sometimes considered a challenge by non-neurologists. A neurologic exam which tests all aspects of all neurologic modalities can quite literally take several hours to perform. On the other hand, a very good, thorough neurologic exam which yields substantial relevant information can be performed in under 5 min. Adherence to a systematic framework or approach to the examination, appropriate for the age and abilities of the child, can be extremely helpful in simplifying the basic questions: Is this child normal, and if not, why not, and how do I describe it? This chapter will focus on how to perform some of the routine elements of the neurologic examination with tips on how to tailor the exam for various age groups.

A.S. May, M.D.

Department of Pediatrics, New York Presbyterian Hospital, 525 E. 68TH ST BOX 91, New York, NY 10021, USA

S.T. Keros, M.D., Ph.D. (✉)

Pediatric Neurology, Weill Cornell Medical College, 525 E. 68th St, Box 91, New York, NY 10021, USA

Sanford Children's Hospital, University of South Dakota Sanford School of Medicine, Sioux Falls, SD 57105, USA

e-mail: Sok2005@med.cornell.edu

The History

As always, obtaining a solid history of present illness is essential for directing the physical exam and establishing a diagnosis, and one is often able to narrow the differential diagnosis substantially based on history alone. As most people who work with children have already learned, simple observation of the child while obtaining the history can also influence one's approach toward obtaining information.

Birth History

It is likely not necessary to remind anyone with a background in pediatrics that any history in children should include details of the pregnancy and birth. Many neurologic disorders, whether genetic or acquired, can begin in pregnancy. Complications in pregnancy, such as growth restriction, failure to progress, fetal distress, or prolonged labor, may indicate antenatal disorders rather than any process which began as a result of the actual birth process itself. One should evaluate for maternal antenatal factors such as illnesses and teratogenic exposures. Of course, the gestational age at which the child was born will also help establish a developmentally appropriate norm. If known, Apgar scores may also be informative and give a general clue as to the onset or timing of childhood disorders.

Developmental History

Typically, developmental milestones are divided into three broad domains—motor, language, and social. Assessing the current developmental milestones achieved is essential in evaluating mental status and cognition. Knowing when some key milestones, such as first words or first steps, were reached will help to classify the severity of any delays. Determining the pattern and timing of any developmental problem is necessary to classify it as static (such as might be seen in cerebral palsy), progressive (as might be seen in some mitochondrial disorders), or regressive (as seen in Rett syndrome, some leukodystrophies and Landau-Kleffner syndrome, to list a few examples). Additional considerations in assessing developmental norms are discussed in the “Mental Status” section.

The General Exam

Although clichéd, it bears repeating that the majority of the neurological examination can be performed simply by paying careful attention to a child’s affect, behaviors, and natural or spontaneous movements. Of course, this does not obviate the need for a systematic, methodical approach to a complete neurologic exam when the situation warrants. There are many neurologic disorders that have systemic “non-neurologic” involvement which are evident during the general physical exam. This chapter will first point out a few key considerations to note while performing a general exam, which are relevant to neurologic disorders, and then proceed to address the conventional neurologic examination.

The Head

The first assessment of the head is to determine its size. This measurement is a way to evaluate the status of the central nervous system in the newborn and early childhood period, because head size is a proxy for the overall volume of the brain

and the cerebrospinal fluid. To measure the head circumference, a tape measure is placed on the occiput posteriorly and placed above the eyebrows anteriorly or along the largest protrusion of the forehead. Ideally this measurement is taken a few times and averaged, as it is can be difficult to accurately measure the head circumference, and there is often variation between measurements. Circumference measurements should be plotted on an appropriate growth chart. Macrocephaly is typically defined as an occipitofrontal head circumference greater than the 97th percentile for age, while microcephaly refers to measurements less than the 2nd or 3rd percentile.

It is critical to remember that any rapid changes or progressive trends in percentile are likely more important than any absolute value at a single point in time.

Rapid increases in head circumference may be the first indication of hydrocephalus. On the other hand, rates of head growth which do not keep pace with weight or length may represent a brain which is not growing as expected. In cases of systemic disease such as malnourishment, head size is relatively preserved compared to length and weight. Please refer to Chap. 14 for a full discussion of the evaluation of a large head.

Next, the anterior and posterior fontanelles should be assessed. The anterior fontanelles should be evaluated with the child in an upright position. An anterior fontanelle which is bulging or firm can be a sign of increased intracranial pressure. The anterior fontanelle typically closes between 7 to 19 months of age, and several conditions are associated with early or delayed closure. However, early or late fontanelle closure without other abnormalities in the exam is rarely a cause of concern. The posterior fontanelle can be closed as early as birth and otherwise closes by 2 months of age.

The cranial sutures, the edges of the bone plates that form the skull, should also be assessed while examining the head. Craniosynostosis, the premature closure of these sutures, will affect the size and shape of the head. Plagiocephaly (literally means “oblique head”) refers to a flattening of a portion of the skull. The most commonly

seen form is positional plagiocephaly, where the occipital region is flattened (usually toward the lateral side) and the ipsilateral frontal area is prominent due to forward protrusion. The incidence of positional plagiocephaly has increased as a result of the “back-to-sleep” campaign. Positional plagiocephaly is primarily a cosmetic issue, which does not affect brain development. Craniosynostosis and plagiocephaly are discussed in more detail in Chap. 6.

The Face

The presence of dysmorphic facial features, if any, may be suggestive of genetic syndromes. While 15 % of normal newborns may have one dysmorphic feature, the presence of two or more of such features is much less common and is associated with increased risk of a clinically significant anomaly. Some constellations of facial features are readily identified by most people (e.g., the classic facies in Down syndrome), but many abnormal facial features can be subtle and not easily recognized. It is helpful to systematically examine and even measure several facial features, as there are well-established norms for comparison. Typically, this type of detailed facial analysis is performed by geneticists. Interestingly, computer automated-dysmorphometry is a growing field of research, and there are case reports where this technique has revealed a genetic diagnosis.

The Neck

The neck should be examined to assess for full range of motion and absence of rigidity or asymmetry, particularly if there are abnormalities of the head or head shape. Infant torticollis, which results in twisting of the head due to a shortened sternocleidomastoid muscle, is a common cause of positional plagiocephaly. Nuchal rigidity is of course a concerning sign which may signify meningeal irritation due to infection or other causes and should be assessed for in any ill appearing child, keeping in mind that nuchal

rigidity is often not present in cases of meningitis, particularly in children.

Cardiovascular and Abdomen

Auscultation of the heart or other large vessels such as the carotid arteries, descending aorta, or renal arteries can reveal murmurs or bruits which may be a risk factor for stroke from causes such as embolism, renovascular hypertension arising from fibromuscular dysplasia, or Takayasu arteritis. Organomegaly, specifically hepatosplenomegaly, can be present in many storage diseases.

The Skin

The skin and the central nervous system both develop from the ectoderm during embryogenesis, and many neurologic diseases are associated with dermatologic findings. There are several classic neurocutaneous disorders where the diagnosis can often be made simply from skin findings. Café au lait spots, axillary or inguinal freckling, and neurofibromas are markers of neurofibromatosis Type 1. Ash leaf spots or hypopigmented patches, adenoma sebaceum, and shagreen patches are lesions seen in children with tuberous sclerosis. Cutaneous vascular lesions, such as capillary malformations involving the ophthalmic region of the trigeminal nerve, are associated with Sturge-Weber syndrome. Additional information on neurocutaneous disorders is provided in Chap. 7.

The Spine and Back

Abnormalities noted in the back or spine, such as discolorations, dimples, or tufts, may be clues to an underlying problem with the spinal cord. Alternately, purely bony problems with the spine, such as hemivertebrae, can cause secondary spinal cord injuries through severe scoliosis or narrowing of the spine. Please see Chaps. 8, 9, and 19 for more thorough discussion of the back, spine, and spinal cord disorders.

The Neurologic Exam

The neurological exam is often presented in a “head to toe” format. With infants and children, of course, the examiner does not always have the luxury of focused patient cooperation as he or she marches down a preset list of areas and functions to be examined. As always, first perform whichever parts of the examination the patient will readily allow. One strategy is to initially assess for tone, range of motion, and deep tendon reflexes, while a child is resting or otherwise calm. If at any point there is crying or screaming, this is a good time to assess the cranial nerves by observing for facial strength and symmetry and palate elevation, for example. Also, an uncooperative child’s limb strength is usually easily assessed by the vigorousness with which the child attempts to evade or terminate the exam.

Mental Status

The mental status and its examination are obviously dependent on the child’s age. By about age 6, one can generally assess mental status and cognition using a similar approach as used in adults. For completeness, a brief review of the mental status for older children will be described. The key components of mental status include the level of arousal, orientation, attention, memory, language, and higher cognitive functions. The level of arousal is typically graded or classified (from best to worst) as awake and alert, drowsy, confused, lethargic, obtunded, stuporous, or comatose. Orientation states range from fully oriented (knowing the date, location, the people in the room) to complete confusion. Standard definitions of orientation do not apply to infants and toddlers, of course. Memory, which includes both short-term and long-term memory, can be assessed with a standard test of recall of named objects after a few minutes or asking about activities performed the day before or perhaps the previous summer. Attention is commonly assessed with a digit span test. Give the child a list of digits to repeat verbally or to dial on a telephone

number pad. Many 4- or 5-year-old children can remember and instantly repeat at least four digits. Basic language skills should also be evaluated, including investigation of receptive language, for example, understanding and following commands, and expressive language, such as spontaneous and provoked speech, naming, repetition, etc. Reading and writing skills are other components of a language examination which can be tested in school-age children.

In neonates and premature babies, the mental status exam is essentially restricted to evaluating for gestational-age-appropriate level of alertness and “general movements,” which describe the highly stereotypical patterns of motions which neonates will do while awake. Before 28 weeks’ gestational age, it is very difficult to note discrete times of wakefulness. By about 28 weeks, one can observe that a gentle movement will cause arousals in the infant. At 32 weeks, eyes begin to open spontaneously and may stay open for extended periods of time. Around 36 weeks one begins to see progressively increased periods of wakefulness and alertness, with vigorous crying. By term, there should be clear attention to visual and auditory stimuli.

In infants between 1 and 6 months of age, the mental status is still primarily an evaluation of alertness and attentiveness, but which now can be also assessed with the motor behaviors which make up the traditional milestones commonly assessed in children. For example, facial tracking requires sustained wakefulness and sustained attention, in addition to an intact visual system and functioning cranial nerves. Thus, in the younger children, the developmental milestones do not neatly fit into any one category of a neurologic exam and blur the lines between mental status, cognition, and motor component.

Cranial Nerves

Although the 12 cranial nerves can be individually evaluated in an order child upon request, in younger children these are often evaluated with observation of spontaneous or provoked responses.

I: Olfactory Nerve

This nerve mediates the sense of smell. This nerve is rarely tested even in adults and has little utility for testing in a child unless a deficit is somehow otherwise suspected.

II: The Optic Nerve

CN II transmits visual inputs from the retina to the areas of the brain which control reflexive movements and unconscious perception (i.e., pupillary light reflexes, blink to threat, tracking and pursuit mechanisms) as well as the conscious perception of light.

The most direct evaluation of the optic nerve is to assess for pupillary light reflex, which requires the proper functioning of the optic nerve to transmit the light information which hits the retina and cranial nerve III which mediates the pupillary constriction. The direct response describes ipsilateral constriction of the pupil, while the consensual response is the constriction which also occurs in the contralateral eye. Pupillary light reflexes should be apparent by 32–35 weeks' gestational age but can be difficult to assess in infants because the pupils are relatively small relative to the size of the iris at this age.

By term, infants should be able to fixate and follow with their eyes, and this tracking response strengthens in the first 2–3 months of life.

The ideal subject to test for tracking is a human face held 8–12 in from the child. An easy way to test for tracking is to hold the child in your outstretched arms, facing you, as you rotate him or her around. It is important to note that tracking at this age is likely due to involuntary responses and deep brain structures and does not necessarily involve higher cortical areas such as primary visual cortex.

Visual field evaluation is not classically a test of optic nerve integrity, although it is reasonable to include a discussion of visual fields in this section. Visual field testing may identify focal retinal deficits as well as brain lesions involving any part of the visual pathway. In cooperative children (and adults), the best method is to position yourself at arms' length away from the patient

and place your palms with fingers extended toward the patient in the area of the visual field you wish to test. Ask the child to look at your nose and to point or look if he or she sees the movement when you briefly flex or wiggle just your index finger a few times. This method of visual field testing is preferred to finger counting or bringing in wiggling fingers from the periphery, as many children will reflexively look toward the end of any outstretched arm and thus requires repeated attempts and urging not to look to the sides and anticipate the stimulus. Testing four quadrants in each eye is usually sufficient, remembering that visual field deficits are not necessarily peripheral and can also be patchy.

In younger children who cannot participate in confrontational visual field testing, an effective method is to hold an interesting object, such as a colorful toy, with both hands behind the examiners head. Then, simultaneously bring the object out into one quadrant of the visual field and an empty hand in the opposing quadrant. If the child can see the object, then he or she will look toward the more interesting stimulus, in this case the toy. In the youngest children, a “blink-to-threat” technique can be used. To do this the examiner brings an object such as a fist rapidly toward the eye. This is repeated, coming from different directions while looking for a blink response. Note that this test can frequently be unreliable, as air movements from an approaching hand or other object may stimulate a blink response. In addition, failure to blink is not necessarily an indication of lack of sight, so one must use caution in over-interpreting the results of blink-to-threat testing.

Finally, the fundoscopic exam is a method which directly visualizes the retinal portion of the optic nerve. This can be very challenging in children and a discussion of fundoscopy is beyond the scope of this chapter. If any visual deficits are suspected in a child, a referral to a pediatric ophthalmologist or neuro-ophthalmologist is suggested for a detailed examination and dilated fundoscopy. Chapter 12 will give an in-depth description of the ophthalmologic exam in children.

III/IV/VI: The Oculomotor Nerve, the Trochlear Nerve, and the Abducens Nerve

The oculomotor nerve, the trochlear nerve, and the abducens nerve are responsible for the movement of the eyes. The oculomotor nerve contains the fibers which mediate the efferent papillary reflex and elevate the eyelid, in addition to being the nerve which innervates most of the muscles which control eye movements (the superior rectus, inferior rectus, medial rectus, and inferior oblique). The trochlear nerve controls the superior oblique muscle, which has a complicated function which varies depending on the direction of gaze, but is essentially responsible for intorting and depressing the eye, while the abducens nerve controls the lateral rectus which abducts the eye for lateral movements.

To formally test for the integrity of these nerves and the associated muscles, assess for full range of motion in the horizontal and vertical directions. Testing the “diagonals” increases the sensitivity of noting any deficits, because the extraocular muscles do not attach to the globe at perfect right angles. If limited range of motion or disconjugate gaze is noted in any direction, each eye can then be evaluated separately in an attempt to determine which eye is the abnormal one. In older children, inquiring about diplopia is important, as patient self-reporting can be more sensitive than directional testing. In infants, use a strong stimulus such as a toy or your own face in an attempt to have them follow, while you assess eye movements. In newborns, using the “doll’s eye” reflex can be used to assess horizontal eye movements. To perform this test, also known as the oculocephalic reflex, the head is turned somewhat quickly but gently to one side. The movement should result in a temporary deviation of both eyes in a direction opposite to the direction of turning. The doll’s reflex is typically present as early as 25 weeks of gestation.

There are several clinically important eye abnormalities that are worth noting. Weakness of the superior oblique muscle or trochlear nerve will result in a compensatory head tilt in many children, in order to prevent the diplopia which results from the abnormal elevation (hypertropia)

of the affected eye. The most common cause of trochlear nerve damage is head trauma. Incomplete abduction of an eye is usually due to weakness of the lateral rectus which is innervated by the abducens nerve. One cause of abducens injury is increased intracranial pressure due to brain edema or hydrocephalus. Also, Duane syndrome is a form of congenital abducens nerve malfunction (occasionally also involving other cranial nerves), which limits eye mobility but is typically benign and does not result in overt visual deficits. Horner’s syndrome is a constellation of findings, which include miosis (pupillary constriction), ptosis, and anhidrosis on the same side of the face, and is frequently caused by disruption of sympathetic innervation which ascends along the carotid artery. However, Horner’s syndrome can also arise from a central brain or spinal disorder.

Parinaud syndrome results in an impairment of upward gaze, often accompanied by eyelid retraction and pupillary abnormalities. Parinaud syndrome is the result of a lesion or compression of the pretectal area in the dorsal midbrain, which is a common location for pediatric neoplasms. Other causes of this syndrome include obstructive hydrocephalus or direct injury due to ischemia or hemorrhage.

V: The Trigeminal Nerve

The trigeminal nerve controls sensation of the face as well as the muscles of mastication. Facial sensation in older children can be tested by applying a stimulus such as light touch to each division of the trigeminal nerve: the ophthalmic branch of the forehead, the maxillary branch (the cheek), and the mandibular branch (the chin). In infants, tickling or stroking the face, for example, with a cotton swab on one side of the nose, cheek, or lip, should elicit a rooting-like motor response toward the side of the face that was stimulated. The trigeminal nerve can also be tested by eliciting the corneal reflex. A very light touch to the cornea, such as with a wisp of cotton, should trigger a bilateral blink reflex. The sensation is mediated by the ophthalmic branch, while the motor response arrives via the facial nerve.

Chewing movements are mediated by the muscles of mastication which are innervated by

the mandibular branch of V. Opening the jaw is due to the action of the external pterygoids while closing the jaw is due to the masseter and temporalis. In neonates the muscles of mastication can be tested indirectly by evaluation of sucking strength and control and more directly by allowing the infant to bite on your fingers.

VII: The Facial Nerve

The facial nerve controls the muscles of facial expression and also mediates taste on the anterior two thirds of the tongue. In cooperative children, facial nerve integrity can be demonstrated by noting strong eye closure, wrinkling of the eyebrows, smiling, and strong puffing out of the cheeks. The face at rest can also be examined to evaluate for any asymmetry such as widening of the palpebral fissure or flattening of the nasolabial fold which might suggest weakness, keeping in mind of course that some slight asymmetries may be normal.

In infants and neonates, evaluating for facial symmetry at rest and while crying or smiling is usually sufficient. Taste in older children can be tested by using a concentrated solution of salt or sugar, placed on the tongue with a cotton swab, and should be reported before putting the tongue back in the mouth to avoid detection with cranial nerve IX. Testing taste is usually done in order to help determine if a facial nerve deficit is due to problems with the nerve (i.e., a “peripheral 7th” such as due to a Bell’s palsy, in which case taste will be impaired) from a “central 7th,” where taste would be expected to be preserved.

VIII: The Vestibulocochlear Nerve

The vestibulocochlear nerve mediates hearing and vestibular function. Finger rubs or whispers can test for hearing acuity. If a deficit is observed, use a tuning fork (256 Hz) to perform the Weber and Rinne tests. In the Weber test, the tuning fork is placed at the vertex of the head. The sound should appear to come from the midline if hearing is intact. However, if one ear is abnormal, the sound will lateralize to the side of decreased hearing in cases of conductive hearing loss (i.e., a problem with the outer ear, eardrum, or ossicles) but will lateralize to the normal ear in cases of

sensorineural hearing loss arising from damage to the cochlea or eighth nerve. The Rinne test is performed by holding the base of the vibrating tuning fork against the mastoid process of the abnormal ear. When the subject no longer perceives sound, the vibrating end of the tuning fork is then placed just outside the ear canal. If the tuning fork can now be heard, this signifies a “positive test” and suggests sensorineural hearing loss. If the tuning fork cannot be heard after removing it from the mastoid, the test is negative and suggests a conductive hearing loss. Note that in normal ears without hearing loss, the normal result is “positive.”

Children begin to clearly localize sound at about 6 months. Use a noisy toy or loud voice while observing patient reaction to evaluate hearing. In younger infants and newborns, a startle response or eye blink to loud or sudden sounds can evaluate for basic hearing ability. This reflex is present as early as 28 weeks. Vestibular dysfunction can manifest with diverse symptoms such as vertigo, nystagmus, emesis, and ataxic movements. Direct tests of vestibular function are beyond the scope of the basic neurological exam and this chapter.

IX and X: The Glossopharyngeal Nerve and Vagus Nerves

The glossopharyngeal nerve and vagus nerves are often tested together. The glossopharyngeal nerve mediates taste and sensation to the pharynx, and the vagus nerve is responsible for many functions, among which is pharyngeal constriction, palate elevation, and vocal cord movement. Symmetric palate elevation while saying “ahh” tests the integrity of the vagus nerve, while the gag reflex tests both the sensory function of the glossopharyngeal nerve and motor aspects of the vagus. Normal swallowing suggests that these nerves are intact, as they must work together for the proper coordination needed to swallow.

XI: The Spinal Accessory Nerve

The spinal accessory nerve innervates the sternocleidomastoid and the trapezius muscles. Head turning against resistance tests the sternocleidomastoid, and a strong shoulder shrug tests the

trapezius muscles. In newborns, observe for proper neck strength with turning of the head. Isolated spinal accessory nerve lesions are rare, however, and routine testing in infants is rarely helpful unless there are other neurologic signs.

XII: The Hypoglossal Nerve

The hypoglossal nerve innervates the tongue, and a deficit will produce deviation to the abnormal side upon tongue protrusion. Tongue strength can also be tested by asking the child to move the tongue from side to side and also to push against the interior of the cheek with resistance applied from the outside the mouth. In any case of suspected nerve or muscle disease, or in infants with generalized weakness (e.g., in suspected spinal muscular atrophy), the tongue should be examined at rest while in the mouth for fasciculations. Tongue fasciculation can be an early indicator of neuromuscular disease. Be careful, however, as a protruding tongue will often have a normal tremor or movement that can be mistaken for fasciculations. In newborns the sucking reflex can be utilized to evaluate lingual tone and strength.

Motor Examination

There are several elements to the motor or strength portion of the neurological exam. First, a simple observation of positioning at rest may show atypical postures or asymmetries which may suggest weakness or abnormalities of tone. The preterm infant, for example, keeps the limbs in an extended position but the normal full-term infant has flexed extremities at rest. Some characteristic poses, such as “frog-leg posturing,” or external rotation of the legs suggests a systemic weakness. Scissoring of the legs, where the legs tend to cross at the feet or ankles, is one sign of possibly increased tone of the hip adductors and is often seen in children with spasticity from upper motor neuron injuries such as corticospinal tract injuries. Fisting of the hand or holding the thumb adducted across the palm is a position which also suggests corticospinal tract involvement. Observation at rest will also usually reveal

adventitious movements such as tremors, tics, and myoclonus.

Another component of the motor exam is tone, which describes the basic resting resistance of a muscle or group of muscles.

Tone is best examined by assessing resistance to passive movement with the subject at rest.

Low tone or hypotonia can be focal, axial (i.e., central or truncal), appendicular (in the limbs), or generalized. Increased tone or hypertonia is generally either focal or generalized.

Hypertonia can be classified as spastic, where the greater the velocity of passive movement the higher the observed tone, or rigid, where the tone is uniform and not velocity dependent.

The assessment of tone can be somewhat subjective and can be influenced by the state of the patient. A resting or sleeping child can have lower tone than while awake. In general, the normal flexed posture of a newborn and relatively increased tone decreases with age until about 6 months and then plateaus.

In newborns, there are certain generalities which can help to describe or indicate abnormal tone. For example, when pulling a newborn’s arm gently across the chest toward the opposite shoulder, there should be increasing resistance felt as the elbow approaches the midline. If the elbow crosses the midline without excessive force, this is an indication of decreased tone, and this is sometimes referred to as a positive “scarf sign.” In infants, axial tone can be evaluated by holding the patient in a ventral suspension and observing the position of the child draped over the arm. Infants with normal tone should make some attempt to lift the head and keep the back arched. Additionally, one can raise the infant from supine to sitting by pulling on the arms and observing for any abnormal head lag (no lag by 6 months of age), a sign of axial hypotonia and neck weakness.

Formal strength testing usually involves testing for strength in an isolated muscle group. Both muscle bulk and strength should be graded. The most common grading system ranges from 0 to 5, where zero corresponds to no movement, 1 represents a flicker of movement or muscle contraction which does not result in limb motion, 2 indicates

movement but not against gravity (i.e., in the plane of the bed), 3 is movement against gravity but without resistance, 4 is movement against resistance, and 5 is full power or movement against strong external resistance.

In infants and young children, strength testing is typically performed while observing functional movements. For example, symmetric and age appropriate reaching, crawling, or cruising suggest normal strength. Of note, asymmetry in reaching or demonstration of a hand preference before about 1 year of age may be a sign of pathologic weakness in the non-preferred arm or hand.

Sensory Examination

The sensory examination includes evaluation of the primary modalities of pain, temperature, touch, proprioception, and vibration sense. Formal sensory testing can be done typically after 5–6 years of age. A complete sensory examination can take a long time to perform in a healthy subject. *In reality, the sensory exam is usually tailored to a specific area of concern where a deficit is already suspected, as it will seldom reveal an abnormality which has not been previously self-reported.* Pain sensation can be tested with a sterilized pin, being careful to apply the same pressure in all areas. However, it is quite difficult to apply consistent, nontraumatic pressure with each application. Furthermore, certain areas of the skin are naturally more sensitive than others. Thus, variations in the exam are to be expected, and mild sensory deficits or asymmetries should be treated suspiciously and placed in context of the overall history and other examination findings. In general, asking the subject if the pin feels sharp is acceptable, as opposed to attempting to quantify the degree of pain or sharpness. Temperature sensation is transmitted together with pain by the spinothalamic tract and can be tested by placing a cool metal object against the skin. Touch is evaluated by using a cotton swab or the examiners finger and gently pressing, not wiping, against the skin. With eyes closed, the subject should be able to identify the location of the stimulus.

Proprioception, or joint position sense, can be tested by holding the sides of a joint (e.g., the lateral aspects of the big toe) and moving the distal aspect of the joint gently and briefly in an up or down direction. A healthy person should be able to detect even very small movements and specify the direction. Vibration sense, which is transmitted together with proprioception in the dorsal column system of the spinal cord, is best tested by placing a 128 Hz tuning fork on a bony portion of a finger, toe, or other joint.

Sensory testing in newborns and infants is usually limited to basic testing while assessing for motor output as a potential response to a stimulus. For example, an infant will usually withdraw a limb in response to a tuning fork. This procedure also provides temperature and touch stimulation and trial-to-trial variability in infant testing is common. Noxious stimuli such as nail bed pressure, pinching of the skin, or pinprick should also elicit a cry or withdrawal. Light touch or tickling will also usually precipitate a withdrawal of the foot, for example. Again, a sensory exam is most sensitive when there is a specific area of concern for neurologic injury.

Reflex Examination

In neurology, reflexes can refer to either the deep tendon reflexes mediated by stretch receptors in the joints and muscles or to developmental and behavioral reflexes. Deep tendon reflexes test the sensory integrity of the stretch receptors and associated sensory nerves and also require a functioning muscle to produce the motor response. Most reflexes are mediated by only one or two spinal nerve roots and are very helpful when attempting to localize an injury. Upper motor neuron injuries (those arising in the cortex of corticospinal track) will lead to a condition where deep tendon reflexes (and overall tone) are elevated, while lower motor neuron injuries (i.e., the cell bodies of the motor neurons in the spinal cord and their associated nerve fibers) or muscular injury will cause depressed or absent reflexes. When testing the tendon reflexes at a joint such

as the elbow or knee, the joint should be relaxed and bent approximately 90°. A brisk tap with a reflex hammer provides the stretch response and the reaction is a contraction of the muscle. The magnitude of tendon reflexes is dependent on the location as well as strength of the tap with the hammer. Tendon reflexes are most commonly tested in the arms at the biceps (mediated by C5 and C6) just anterior to the elbow, the brachioradialis (also C5/6) above the wrist on the radial aspect of the forearm, and the triceps (C6/7) just posterior to the elbow. In the legs these include the patellar reflex (L2/L3/L4) elicited by a strike just below the patella and the ankle reflex (L5/S1/S2) produced by tapping the Achilles tendon, usually while maximally flexed. The adductor reflex (L2–4) is tested by tapping the adductor tendon near the medial epicondyle of the distal femur. The most common grading system ranges from 0 to “4+” where 0 refers to no reflex, 1+ is a weak reflex, 2+ is a normal reflex, 3+ is hyperactive, and 4+ generally indicates some clonus or spreading. A “crossed adductor response” is one of the more common examples of a reflex spreading and occurs when a tap of one thigh adductor leads to bilateral adductor muscle contraction. Clonus is most easily assessed at the ankle, elicited with a rapid jerk of the ankle into a dorsiflexed position. If present, clonus will appear as a repeated movement or beating of the dorsiflexed foot into plantar flexion. Some beats of clonus can be normal in newborns and young infants as well as teenagers and some adults, but many beats of clonus or sustained clonus should prompt a more thorough examination of possible upper motor nerve injury.

Although deep tendon reflexes can be tested at any age, the triceps reflex is particularly difficult to elicit in neonates. Tendon reflexes can also be difficult to achieve in babies because of their constant motion and light weight, and thus nonspecific movements as well as movement due to the hammer itself can be interpreted as a reflex response. In addition, their small size makes for a small target; the biceps tendon and patellar tendon require a precise hit from the hammer, which can take practice or repeated trials. With children, it is often sufficient to simply use “low” or “brisk”

and then allow the examiner to classify the reflexes as abnormal or normal depending on the rest of the exam.

There are also some reflexes known as “non-stretch” or superficial reflexes. Among these is the abdominal reflex which is elicited by stroking or lightly scratching one side of the abdomen. This should trigger a small ipsilateral contraction of the abdominal muscles and is one way to test for sensation and motor integrity of the thoracic roots 8 through 12, which may be damaged in a spinal cord injury, but does not otherwise have large associated muscle groups for easy testing. The cremasteric reflex causes elevation of the ipsilateral testicle upon upward stimulation of the inner thigh. This reflex is mediated by L1 and L2. Anal tone and the anal “wink” reflex are mediated by S2–S4.

One of the most widely tested superficial reflexes is the plantar reflex.

In children aged 1 or older, a normal plantar reflex produces a plantar flexion of the large toe, while an upward or extensor response is considered abnormal. An extensor response of the toe in non-infant is evidence that this otherwise spinally mediated reflex is not suppressed by information from the cortex.

Thus, an extensor response is another sign of upper motor neuron injury. There are nearly a dozen methods which can be used to test for the plantar response, but the most often used is Babinski test. The Babinski test involves stroking the lateral side of the bottom of the foot up toward the ball of the foot and then curving in toward the toes. An initial upward movement of the big toe is considered a “positive” Babinski sign. A downward movement or no movement is considered the normal situation. The plantar reflex is mediated by the L4–S2 spinal roots. In some cases, a plantar or extensor response is unequivocal. However, testing can be variable from trial to trial, particularly because it is sometimes difficult not to elicit a tickle or withdrawal response upon testing. Note that infants will have a normally positive Babinski sign, as a result of an immature brain. However, a lack of a Babinski sign in an otherwise healthy infant does not have particular neurologic significance.

Developmental Reflexes

These are the final group of reflexes which are commonly tested as part of the neurologic exam. These are reflexes typically present early in life that extinguish with progressive maturation of cortical inhibitory function. These reflexes have two useful functions in neurology. First, their persistence past a certain age may be a clue of a systemic problem. Secondly, and somewhat more commonly, is that these reflexes can be used to elicit a motor response which an infant would otherwise not do voluntarily.

For example, the Moro reflex can be valuable in trying to determine whether there is asymmetric weakness or injury such as might occur in a clavicular fracture or perhaps a brachial plexus injury during birth. The Moro reflex refers to the abduction and subsequent adduction of the arms elicited by a sudden loss of head or trunk support, which creates a sensation of falling. This reflex is readily elicited in a newborn and disappears by 5 or so months of age. *A grasp response is the well-known curling of the fingers or toes when a finger or other object is placed in the palm of the hands or against the balls or soles of the feet.* The sucking reflex and rooting reflexes are also examples of common developmental reflexes and can be used to stimulate motor activity for neurologic testing. The stepping reflex is present during the first 6 weeks of life. This reflex is stimulated by holding an infant upright and resting the feet against a surface with the knees slightly bent. The typical response is for the infant to raise a leg

and move it forward, as if taking a step. The Landau response is triggered with ventral suspension of the child or placement in the prone position, which should result in an extension of both the head and legs. This reflex begins around 3 months and persists through infancy. The tonic neck response describes a flexion of the limbs of the contralateral side and extension of the limbs on the ipsilateral side in response to a force head turn. This usually diminishes by 4–5 months, which is the age many babies begin to roll over. Of note, a normal tonic neck reflex can provoke asymmetries in tone and position, and thus it is important to make sure the head is midline during a neurologic examination. Refer to Table 2.1 for a list of some developmental reflexes and their expected age resolution.

Coordination

Although coordination is commonly thought of as cerebellar-mediated process, coordination is the integration of many different pathways which are all necessary to produce accurate movements. As a result, impaired coordination can be seen in disorders of the cerebellum and basal ganglia as well as various other sensory and motor systems. Finger-to-nose testing is an easy way to assess coordination and simply requires the subject to move his or her finger from the nose to an examiner’s finger or other object, which should be placed as far from the subject while still being reachable, as this distance adds some sensitivity

Table 2.1 Developmental reflexes and their expected age of resolution

Reflex	Stimulus	Response	Resolution
Rooting	Stroke cheek or lip	Head turns and mouth opens toward stimulus	2–3 months
Stepping	Hold upright with feet touching surface	Moves feet as if taking steps	3–4 months
Tonic neck	Placed on back, turn head to side	Ipsilateral arm extension and contralateral flexion	4–5 months
Moro	Sudden falling motion or loud noise	Arms extend and abduct, then adduct	5–6 months
Palmar grasp	Press palm surface	Finger curl	5–6 months
Plantar grasp	Press sole of foot	Toes curling	9–10 months
Landau response	Place on stomach	Back arches and head raises	Starts at 3 months, resolves at 12 months

to the test. The subject should be asked to go from finger to nose and back somewhat briskly, but excessive speed is not necessary to evaluate coordination. Rather, the subject should be able to accurately and very precisely touch the fingertip to the examiner's fingertip. During this test, observe for any dysmetria, which can take the form of missing the target laterally or "pass pointing." Beware of any vision abnormality or impaired depth perception which will confound the results. Finger-to-nose testing is also a good way to evaluate for an intention tremor. An analogous test in the lower extremities involves having the patient place the heel of one foot on the knee of the opposing leg and carefully sliding the heel down the shin toward the ankle and back again. Another test of coordination is the rapid alternating movements test, which when abnormal is called dysdiadokinesis. One way to perform this test is to have the subject rapidly tap the index finger to the thumb or alternately to tap a thigh or other surface with the dorsum of the hand and then the palm of the hand as quickly as possible. Rapid foot tapping is an analogous test in the lower extremities. Note that mild weakness will tend to slow any rapid alternating movements, and thus this is a good test for evaluating subtle unilateral weakness in a hand or foot, by comparing speed of movements to the other side.

Testing coordination is not feasible in newborns and infants, due to lack of established circuitry, in addition to the obvious inability to follow commands. By several months of age, however, simply testing for abnormal movements upon reaching for objects will give some sense of coordination and must of course be compared to the abilities of other children at that age. Midline or truncal ataxia, which can arise from lesions in the vermis of the cerebellum such as might be seen in a medulloblastoma, will affect the ability to sit normally or will affect the balance of a walking child.

Gait

A gait exam is an essential part of any neurological examination, and a normal gait requires the normal functioning of all components of the

nervous system. To some degree, if a child can successfully walk or run down a hallway, stop, and return when asked, the majority of the neurologic exam is surely normal, as they have demonstrated relatively intact hearing, cognitive, motor, visual, and sensory pathways. It is best to evaluate gait by observing the child while barefoot. Note the station or the stance of the patient, their steps, the speed, and the steadiness of the gait. Steadiness varies with age and should be considered in the context of the patient's age. Cooperative children should be asked to walk regularly, then on their toes and heels, and finally with one foot directly in front of the other. This ability to tandem walk is usually present by 5 years of age. For many abnormalities, the gait will be affected in predictable or stereotypical ways. For example, focal weakness in one leg may make toe walking difficult on that foot or result in a limp or other noted asymmetry. Truncal ataxia or ataxia of a leg will result in a state of imbalance and appear clumsy and staggered, with a wide-based gait. Arm weakness or tone may be seen as a decreased arm swing. Other characteristic gait abnormalities are somewhat less intuitive to use in pinpointing the abnormality. Circumduction, for example, is a lateral swinging movement of the foot with excessive lifting at the hip while bringing the foot to the front, which is seen in cases of foot drop or distal weakness. A spastic gait due to increased tone, such as might be seen in cerebral palsy, will appear shuffling, with the legs adducted and the child somewhat on the toes or front of the foot. An antalgic gait describes the traditional limping pattern seen with maneuvers to limit bearing weight on a painful leg.

Obviously, gait can only be tested once a child begins walking. However, an evaluation of crawling, standing, or cruising can be relatively effective proxies for the information provided by independent walking.

Conclusion

The neurologic examination as presented in this chapter is intended to be a brief reminder or summary of various ways to identify and describe

suspected neurologic abnormalities in children. There is of course tremendous variation among clinicians in their overall approach and examining techniques. However, the examination and its components as described represent a solid framework which should help any pediatric health-care provider when trying to determine whether a particular set of complaints, symptoms, or signs are abnormal and whether they may warrant additional work-up and neurological or neurosurgical consultation.

Clinical Pearls and Red Flags

- While a systematic and complete neurologic exam is occasionally needed, observation alone of a child at rest and play can elucidate a tremendous amount about both normal and abnormal neurologic findings.
- To some degree, if a child can successfully walk or run down a hallway, stop, and return when asked, the majority of the neurologic exam is surely normal, as they have demonstrated relatively intact hearing, cognitive, motor, visual, and sensory pathways.
- Understanding and interpreting the neurologic exam in children requires in-depth knowledge of normal development.
- Handedness in infants under 1 year of age may indicate a relative weakness or neurologic problem in the nondominant side.

Suggested Reading¹

- Aminoff MJ, Simon RR, Greenberg D. Clinical neurology. Europe: McGraw-Hill Education; 2015.
- Fenichel GM. Clinical pediatric neurology: a signs and symptoms approach. 3rd ed. Philadelphia: WB Saunders; 1997.
- Newman SA, Ropper AH, Brown RH. Adams and Victor's principles of neurology. 8th ed. New York: McGraw-Hill Medical; 2005.

¹There are many excellent basic neurology textbooks which contain extensive discussions about the various aspects of the neurologic exam presented in this chapter. Several are listed here in lieu of a specific reference list.

Common Neurosurgical Conditions in the Pediatric
Practice

Recognition and Management

Greenfield, J.P.; Long, C.B. (Eds.)

2017, XV, 444 p. 299 illus., 110 illus. in color., Softcover

ISBN: 978-1-4939-3805-6