The Neurological Examination: Pediatrics

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1. Introduction

The pediatric adage that, "Children are not just small adults," rings true in neurology. While the foundation of pediatric neurology fits well within the framework of classical neurology, there are special considerations in the examination and evaluation of neonates, infants, and children. The clinician should endeavor to utilize the rostro-caudal organization of the neuraxis when presenting the examination, ie, an orderly presentation of mental status, cranial nerves, motor systems, and so forth. Yet the approach to the child must take into particular consideration a nonthreatening, flexible style, requiring a patient demeanor and lighthearted affect. The pediatric neurologic examination relies on the time-honored discipline of neurologic localization, but also accounts for the process of normal development. This chapter will select aspects of the neurologic evaluation that supplement the adult examination and are of special emphasis in pediatrics. Particular key points will be made to aid in practical applications of common findings, although a comprehensive differential diagnosis of features on the examination is beyond the scope of this chapter. As the experienced clinician will often say that the lack of a diagnosis upon the completion of the history invariably means the lack of a diagnosis upon the completion of the physical, we will begin with some notes regarding the pediatric history.

2. Pediatric History

The breadth of pediatrics covers the fetal period to adolescence. Naturally, the history and examination will evolve over the individual's lifespan. The neonatal period includes the first 30 days post-term (called postconceptional age, although postmenstrual age is more accurate). The history of any neonate, infant, or child should include a thorough assessment of the pregnancy, labor, and delivery. The gestational history includes the mother's age, duration of pregnancy, and history of maternal illnesses or fetal exposures to teratogens such as drugs or radiation. The TORCH viral infections, causing fetal cerebral injury (and frequently deafness and ocular problems), refer to toxoplasmosis, syphilis, rubella, cytomegalovirus, herpes simplex, and HIV. The timing of an adverse exposure or illness during pregnancy is critical to the neurologic outcome. The nervous system begins with the process of neurulation, or formation of the neural tube, at 17 days post-conception, and this process continues until approximately 7 weeks post-conception. Ventral induction, or formation of the hemispheres, begins at 5 weeks post-conception; disruptions here may lead to holoprosencephaly, or a single, unilobar cerebrum, or to milder versions along this spectrum. Brain development proceeds with nerve cell proliferation between 2 and 4 months of gestation, then neuronal migration between 3 and 5 months, and then nerve cell organization and the formation of synaptic circuitry. Disorders of proliferation include macrencephaly and microcephaly vera. Migrational disorders include lissencephaly (smooth brain), pachygyria (thick gyri), polymicrogyria, and schizencephaly (congenital clefts). Myelination occurs, starting inferiorly and posteriorly, during the latter half of pregnancy and continues actively until two years post-term. Prenatal encephaloclastic lesions, whether from viral infections or vascular accidents during the fetal period, include porencephaly and hydranencephaly. Knowledge of the orderly process of congenital brain development lends perspective on congenital brain anomalies.

Certain patterns suggest prenatal problems. Maternal bleeding during pregnancy raises concerns regarding pre-existing placental or fetal abnormalities. Toxemia, with maternal edema, proteinuria, and hypertension, is associated with prematurity, placental insufficiency, in-utero growth retardation (IUGR), and neonatal hypoglycemia. Infants of dia-

betic mothers have neonatal hypoglycemia and hypocalcemia, macrosomia, respiratory distress syndrome, and sometimes icterus. Mothers with hypothyroidism or phenylketonuria are at risk of producing offspring with cognitive deficiencies. Mothers with myasthenia gravis may produce infants with neonatal myasthenia, reflecting a transient condition due to passive transfer of acetylcholine-receptor antibody requiring supportive care until resolution. A history of maternal polyhydramnios suggests an infant with a neuromuscular problem who had decreased swallowing. A low maternal alpha-fetoprotein (AFP) suggests Down syndrome, versus high AFP incriminating a neural tube defect.

The perinatal history should include whether the amniotic fluid was clear or meconium stained, and whether the membranes ruptured spontaneously or with medical assistance. Rupture of membranes 24 hours prior to delivery increases the risk of fetal amnionitis, with common organisms being *Streptococcus* Group B, *E coli,* and *Listeria monocytogenes*. Either a prolonged, difficult or precipitous labor suggests problems with the newborn. A fullterm infant is delivered between 37 and 42 weeks of gestation, and on average weighs 2500 grams. Infants small for gestational age are known as SGA or having IUGR, ie, in-utero growth retardation. Infants born large for gestational age (LGA) should raise the question of maternal diabetes, which may have been limited to gestation and unrecognized.

The perinatal history should emphasize whether there was evidence for a newborn encephalopathy, ie, hypotonia, decreased motoric activity, apnea, seizures, temperature instability, or other problems referable to the nervous system. This is the best predictor of whether a neonate is at risk for neurodevelopmental sequelae related to the perinatal period. The APGAR scores, done on a scale of 1 to 10 (worst to best), indicate how well a newborn is doing at that time, but do not have longterm predictive value unless the very late APGAR scores (10 minutes and beyond, as opposed to the standard scores taken at 1 and 5 minutes) are very low. The primary risk factors for the development of cerebral palsy are prematurity and "multiples," ie, twins, triplets, etc., as opposed to birth asphyxia.

The pediatric neurologic history also must concentrate on the developmental history. Each age has its own developmental norms, and premature infants are generally assessed for their "corrected" postconceptional age at least until 12 months. A newborn infant with normal vision should be able to briefly fixate with open eyes. A considerable amount of time, on average 16 hours out of 24, will be spent asleep, with a preponderance of "active" sleep (the forerunner of stage REM) in the first months of life. Screening tests such as the Denver Developmental Screening Test (Denver II) are available to assist with rapid identification of developmental norms and delays. Typical developmental milestones are noted in Table 1. It is important to recognize that there is a wide normative range, and multiple aspects of delay in a particular sphere (gross motor, fine motor, language, social) should be present, often in combination with multiple spheres, before diagnosing a child with a neurodevelopmental disorder.

Children often present with paroxysmal problems, whereupon only the history will provide clues unless the family can produce a videotape of the event. The differential diagnosis of seizures versus nonepileptic paroxysmal phenomena is wide, but special consideration is given to whether there are provocative features as expected in breathholding spells, hypoglycemic events, conversion disorders, etc. The pediatric migraine spectrum includes many "equivalent" syndromes of variable prognoses, eg, benign paroxysmal vertigo, benign intermittent torticollis, periodic syndrome with vomiting, acute confusional migraine, alternating hemiparesis, and others.

The presenting problem must be defined not only by its special characteristics but also by the temporal course. In particular, one must emphasize whether an encephalopathy appears static or nonprogressive versus progressive. The latter is taken to mean the presence of a deteriorating course, where there is regression, ie, loss of previously acquired skills, or a failure to properly acquire new skills. The presence of a progressive or intermittent pattern raises consideration of the inborn errors of metabolism. Such metabolic encephalopathies can be heuristically organized into several patterns. One scheme is to consider whether the disorder is typified by predom-

Table 1

Typical Developmental Milestones

1 month
- Spontaneous, symmetrical motor activity
- Lifts head when prone
- Regards surroundings
- Follows visually to midline

2 months
- Smiles spontaneously and reciprocally
- Coos
- Follows visually past midline

3 months
- Follows 180 degrees
- May wave at toy
- Turns head to sound
- In ventral suspension, head held above body

4 months
- Grasps rattle
- Good head control when placed in sitting position
- Laughs aloud
- Excites with feeding

6 months
- Reaches with either hand
- Rolls over, usually prone-to-supine first
- Sits briefly when placed
- Plays with examiner

7 months
- Transfers objects from hand-to-hand
- Imitates sounds
- Responds to name

8 months
- Sits independently
- Stands holding on
- Combines syllables
- Regards self in mirror

10 months
- Pulls to stand
- Thumb-finger opposition grasp
- Waves bye-bye
- May say "mama" or "dada" (specifically)

12 months
- Walks with support
- Stands alone
- Places cube in cup
- 2 words besides "mama" or "dada"

15 months
- Toddles
- Pats pictures
- Uses spoon
- Points or vocalizes to indicate wants
- 4 to 5 words

18 months
- Removes shoes/socks
- Climbs on furniture
- Stacks 3 to 4 cubes
- 10-word vocabulary
- Turns pages in groups

2 years
- Runs
- Walks up steps one at a time
- Begins to use pronouns, 3- word sentences
- Uses own name
- Toilet trained during day

2-1/2 years
- Partially undresses
- Attempts to put on socks
- Imitates horizontal or vertical line
- Helps put things away
- Knows name and sex

3 years
- Alternates feet upstairs
- Pedals tricycle
- Builds tower of cubes
- Copies a circle
- Speech at least 50% intelligible
- Buttons and unbuttons, puts on shoes

4 years
- Runs and climbs well
- Alternates feet downstairs
- Hops on one foot
- Throws ball overhand
- Copies a cross
- Asks questions
- Imaginative play

5 years
- Skips feet
- Copies a triangle
- Counts to 10 or higher
- Knows age
- Can tie shoelaces

6 years
- Draws person with hands and clothes
- Knows morning and afternoon
- Knows right from left

inantly a leukodystrophy, with white matter involvement, or poliodystrophy, with gray matter involvement. The leukodystrophies typically present with the long tract signs of spasticity, hyperreflexia, and ataxia, as well as visual loss with optic atrophy and deafness. The poliodystrophies will present with more typical gray matter symptoms, such as dementia, seizures, or retinal pathology. As time proceeds, many of the metabolic encephalopathies will share features of both, and clarification of the findings at presentation may help to delineate which broad category is a better fit. Other taxonomic schemes will use classification by organelle, eg, mitochondrial versus peroxisomal. Newer information is permitting the definition of disorders by neurotransmitter metabolism, eg, Segawa DOPA-responsive dystonia with diurnal variation. Clues from the history, along with a knowledge base regarding these conditions, may lead to a well-honed diagnostic plan to verify a suspected diagnosis. Alternatively, less well-defined syndromes may be better evaluated through examinations that allow for screening of multiple potential disorders. The autism spectrum disorder, representing a syndrome that presents commonly to the child neurology clinic, has features of both a static and progressive course, oftentimes with periods of developmental plateau. Thus, the assessment of a temporal course to a child's neurodevelopmental disorder also requires a degree of flexibility that allows for vagaries attributable to the natural history of disease that do not follow the timelines of our diagnostic algorithms.

3. Family History

The family history is of particular importance and should not be dismissed or abrogated to a cursory review. The contributory value of the family history, while traditionally highly honored in child neurology, has become even more amplified with the information explosion in neurogenetics and mapping of the human genome. A family tree represents the optimal pictorial demonstration, and all first degree relatives (parents and siblings) should be represented at the very least, with inquiry into the medical status of grandparents and parents' siblings as well. Disorders of autosomal dominant (involving consecutive generations), autosomal recessive (skips generations), X-linked and mitochondrial (maternal transfer to sons) inheritance may be identified. Parental ancestry and assessment for consanguinity are key points.

4. Physical Examination

The examination is best used to confirm or refute the differential diagnosis as formulated following the history, and should emphasize apropos portions of particular relevance or interest. The pediatric neurology examination requires both the general and neurological examinations. Observing and playing with the child yield high impact information. The general examination in particular should assess for dysmorphic or cutaneous stigmata. Dysmorphic features may involve the cranium, facies, eyes, ears, mouth, skeleton, or extremities. The phenotype may immediately suggest a known syndrome, ranging from the upslanting palpebral fissures of Down syndrome to the shortened palpebral fissures with smooth philtrum and thin upper lip of fetal alcohol syndrome. Coarse facial features suggest the mucopolysaccharidoses.

Neuromuscular disorders may reveal themselves with their own physical signs. The presence of a high-arched palate or pectus excavatum suggests hypotonia of prenatal origin. In a hypotonic newborn, examination of the mother may reveal stigmata of myasthenia to support transient neonatal myasthenia, or myotonia manifest by a delayed handshake release to suggest neonatal myotonic dystrophy. In an older child or adolescent, pes cavus may be present along with chronic peripheral neuropathy as in Charcot-Marie-Tooth syndrome. Assymetric movement of the upper extremities, with persistent forearm extension and pronation in an infant, is seen in Erb congenital brachial plexus palsy. The presence of a Horner syndrome, including heterochromic irides where the ipsilateral eye is lighter in color, indicates additional involvement of the lower plexus as in a complete (Klumpke) palsy.

The phakomatoses, or neurocutaneous disorders, provide a wealth of findings on the general physical examination. The cutaneous findings of tuberous sclerosis include the hypopigmented "ash leaf" maculae, typically lumbar shagreen patch, malar fibroadenomata (previously called adenoma sebaceum), and periungual fibromas around the nailbeds. Typically, the infant in whom the diagnosis is first considered, particularly in the setting of infantile spasms, will only demonstrate the ash leaf spots, which may be better visualized with the aid of a fluorescent (Wood's) lamp. A facial port-wine stain does not always mean there will be cerebrotrigeminal vascular angiomatosis of Sturge Weber syndrome, but involvement of the eyelid and cranial nerve V-1 frontalis distribution is very suggestive. The prepubertal child with neurofibromatosis-1 will demonstrate at least six café au lait maculae greater than 5 mm, along with axillary or inguinal freckling. In incontinentia pigmenti, a female neonate will have transient vesicular lesions sometimes confused with neonatal acne, but which leave linear and whorled hyperpigmented patches seen in older infants and children.

The cranium deserves its own examination. All time-honored modalities of observation, palpation, auscultation, percussion, and transillumination are valuable. A newborn will commonly have some head molding, which resolves within the first weeks of life. Caput succedaneum is self-limited scalp edema. A cephalohematoma represents subperiosteal hemorrhage. Overriding sutures raise concern regarding microcephaly; split sutures and a bulging fontanelle suggest increased intracranial pressure. Anomalous cranial shapes may represent transient molding or a more significant craniosynostosis. The cranium will grow in the axis of the closed suture, so that sagittal synostosis leads to an abnormally elongated head (scaphocephaly), whereas coronal synostosis leads to a shortened, box-like shape (brachycephaly). As parents are now advised to place their children supine during sleep to decrease the risk of sudden infant death syndrome, more and more children present with simple molding, which can be avoided by use of the prone position during waking hours and alternating the child's head position during sleep. Cranial bruits, while often normal in young children, may instead suggest the presence of a vein of Galen malformation, particularly in a young infant with macrocephaly and congestive heart failure.

The posterior fontanelle may be difficult to palpate, and is normally closed at birth, and no later than six weeks. Congenital hypothyroidism should be ruled out in a newborn with an open posterior fontanelle. The anterior fontanelle typically closes between 10 and 20 months of age. When palpated to assess for intracranial pressure, the infant should be sitting up and relaxed. It is normal to perceive the cardiac pulse while palpating the anterior fontanelle. The head circumference measurement should be taken

as the largest consistent frontal-occipital measurement. While normative graphs should be used to calculate the percentile for a male versus female, the general rate of cranial growth is helpful to know at the bedside. A term infant has an average FOC (fronto-occipital circumference) of about 34 cm. The FOC grows rapidly, at a rate of 2 cm per month the first three months, reaching 40 cm at age three months. The rate slows to 1 cm per month the next three months, so that a six-month-old typically has an FOC of 43 cm. The growth is then on average 0.5 cm per month the next six months, or 46 cm at 12 months. Over the next two years, the rate is about 0.5 cm per six months, or 48 cm at 2 years and approximately 50 cm at 3 years. The FOC is about 52 cm at 8 years, and 54-55 cm in the adult. Macrocephaly is often a benign familial trait, but raises concern regarding a plethora of diagnoses, ranging from an intracranial tumor or posterior fossa cyst with hydrocephalus, eg, Dandy-Walker malformation, to subdural hematomata or hydrocephalus that may be related to child abuse.

The general examination may pick up many important findings that may not be manifest by an abnormal neurological examination early in life. A sacral dimple above the gluteal fold, possibly accompanied by a tuft of hair or sinus tract, may be the only clue to spinal dysraphism and a tethered spinal cord that will lead to later neurological deficits during growth. Involvement of other organ systems may provide important support of a potential neurological diagnosis, eg, hepatic or renal dysfunction in a neonate with suspected perinatal hypoxic-ischemic encephalopathy, or thrombocytopenia and hepatosplenomegaly in a neonate with a TORCH infection. A cardiac murmur may accompany the atrioventricular canal defect of Down syndrome, or a ventricular septal defect associated with fetal alcohol syndrome. An abdominal mass in an infant with proptosis is alarming for metastatic neuroblastoma. Clearly the distinction between benign variants and pathognomonic signs on the general examination is critical in pediatric neurology.

Mental Status

The classic mental status examination is obviously adjusted for the patient's age. A responsive and calm, or at least consolable, infant is more reassuring for normalcy than persistent irritability or unresponsiveness. A newborn with an excessive Moro response, or persistent jitteriness, raises concerns regarding neonatal drug withdrawal following maternal drug use, electrolyte disturbance including hypoglycemia or hypocalcemia, or other encephalopathy. An excessive startle reaction on the "nose tap" test is pathognomonic for hyperekplexia. Loss or lack of visual attentiveness is concerning in an infant, and sometimes is the first or only developmental change in infantile spasms. Auditory myoclonus to a startle is worrisome for various encephalopathies and is a well-known feature in Tay-Sachs disease.

The developmental milestones shown in Table 1 are helpful guideposts during the examination. In the preschool or school age child, age and grade relevant questions and even brief tests add considerable strength to the mental status examination. Screens such as the Beery Test of Visual Motor Integration (VMI), Draw A Person test, and WRAT-R (Wide Range Achievement Test - Reading) are readily available and allow for rapid acquisition and scoring. Referral for psychoeducational testing, whether through the school, clinical psychologist, or neuropsychologist may be predicated upon the child's performance. A common presenting problem to the child neurologist is the question of a primary attention disorder, learning disability, or combination of these problems leading to academic or behavioral impairment.

The acquisition of language is of particular importance in the mental status examination. The early milestones are included in Table 1. The speech of a three-year-old should be at least 50% understandable. By this age, a child should also be using at least 2-3 word sentences. As the child becomes older, more correct use of pronouns, verb tense, and noun plurals emerges. The infant begins consonants with reduplicated babble, eg, dada-dada, then nonreduplicated babble, and then moves on to jabbering. The process of articulating the various combinations of consonant sounds occurs between the

ages of 3 and 7 years. Persistent difficulty with specific sounds may be secondary to misaligned dentition, or uncoordinated movements of the tongue and lips. Ongoing dysarticulation, or stuttering characterized by conscious tension and speech avoidance, should lead to referral for speech and language therapy.

Higher cortical functions can be screened during the neurological examination, and may be helpful in the evaluation of a child for the so-called soft or subtle neurologic signs that accompany ADHD and learning disabilities. Sequential complex movements, such as repeatedly tapping the palm to the thigh, then rotating 90 degrees to the ulnar hand surface, and then producing a fist, may demonstrate dyspraxia related to the contralateral prefrontal cortex of motor planning. Testing for finger gnosis may be done by having the child with closed eyes name the finger that is touched by the examiner. Right-left discrimination is learned at about age six years, and same-side commands can be accomplished, ie, "touch your right thumb to your right ear." Cross sided commands are accomplished in the somewhat older child. Right-left discrimination may be impaired in normal individuals, but is also found in association with learning disabilities.

Cranial Nerves

The newborn infant should be able to show some visual fixation as well as reaction to olfactory, gustatory, and auditory stimuli. The facial grimace should be observed carefully. An asymmetric smile could represent congenital facial nerve palsy, although it may also manifest the asymmetric crying facies syndrome secondary to congenital absence of the depressor anguli oris muscle. This occurs in conjunction with a congenital heart defect in up to 20% of cases, and hence is known as the cardiofacial syndrome. The velocardiofacial syndrome may be detected by the combination of a palatal defect and a cardiac murmur; these infants have a telltale facial sign of a very wide nasal bridge. The cranial nerve functions are also examined by the rooting (CN V and VII) and sucking (CN V, VII, and XII) reflexes in the newborn.

Cranial nerve testing in young children often requires considerable adjustment in technique compared to the traditional neurological examination. Simple observation may be sufficient. Torticollis present from birth often reflects sternocleidomastoid muscle spasm of prenatal onset and responds to physical therapy with passive stretching. Acquired torticollis, however, suggests pathology involving the posterior fossa. Head tilt may be seen in the syndrome of spasmus nutans, with asymmetric ocular oscillations and head nodding. Alternatively, a head tilt could represent compensation for a contralateral CN IV palsy, or cerebellar tonsillar herniation through the foramen magnum, that may reflect a posterior fossa lesion. Amaurosis in combination with nonparalytic strabismus can be an unrecognized finding for a number of years. In the young child, visual acuity can be assessed indirectly by determining if the patient reaches for small objects. In a preschooler, one can use the direction of the "E" or three fingers, with the patient responding by showing the direction. If visual acuity appears to be deficient below the level of counting fingers or mimicking direction, a bright light in a dark room may be used to determine for light perception (LP). The ability to tell the direction from which the light is shining is called light perception with projection (LP with P). Opticokinetic nystagmus can be observed by using a large commercially available drum or tapes with bright figures or stripes in young children. The response may be deficient when targets are moved toward the involved hemisphere. The infant too young to follow the rotating drum will normally demonstrate so-called twirling nystagmus with a spinning movement. The oculocephalic, corneal and gag reflexes are present from birth. It is often best to delay the cranial nerve examination until the end of the session, due to its more threatening nature, although one risks the eventual fatigue and uncooperation of the patient.

Motor

Careful observation followed by gentle palpation and passive range of motion are first steps. At term, there is generally more flexor tone. There can be some persistence of clenched fists and internal thumbing until four months of age. In contrast, the premature infant will tend toward hypotonia. The degree of flexor tone

predicts gestational age. Hypo-tonia in the term infant suggests neurological dysfunction, whether related to hemispheric, spinal cord, or lower motor neuron pathology. The most common causes will be central, and include a multitude of systemic problems as well as genetic syndromes and congenital brain malformations. Intracranial hemorrhage or spinal cord injury may produce persistent hypotonia, despite the upper motor neuron localization. A frog-leg posture or reduced motoric activity in a young infant suggests a significant myelopathy or lower motor neuron disorder. Spinal muscular atrophy type I, Werdnig-Hoffman disease, presents within the first six months of life. This patient will typically have a very alert appearance but profound hypotonia and hyporeflexia.

Head and neck control develop over the first several months of life. The head lag as a child is pulled by the hands to a sitting position is resolved normally by 4 months of age. By this age, the child supports some weight with the legs. Sitting is achieved with confidence between 7-8 months, followed typically by creeping, crawling, and then cruising by 10 months. There is a wide normative range in development, especially in the gross motor realm. The significance of "late walking," or absence of crawling, should be judged in the context of associated motor findings. Associated hypotonia does raise the question of "benign congenital hypotonia" or "dysmaturation," which may have a benign outcome but raises the question of a motor system disorder. Motor activities become progressively more purposeful and complex throughout the first year of life, and any persistent asymmetry suggests a neurological problem, including prominent handedness before 9-12 months of age. During the second year, walking improves and becomes confident typically by 18 months, and running begins with reliance on the upper extremities for balance and protection.

Individualized muscle testing is often not evaluable, yet functional activity can be observed in a multitude of ways. The Gower's maneuver, a time-honored test for proximal weakness, is optimally done with the child beginning supine. The weak child will need to roll over to the side before arising, to be followed by climbing up one's own legs with sequential hand movements to reach the standing position.

Reflexes

The traditional reflexes, ie, deep tendon and superficial, are the parlance of traditional neurology and covered in the adult neurological examination. Other than a positive patellar tap, the deep tendon reflexes are difficult to elicit in the newborn. The ankle and triceps reflexes do not appear until several weeks of age, with the triceps being inconsistent until six months. Alternatively, hyperactive responses such as the crossed adductor are typical up to 7 months, and an extensor plantar response is common up to age one year and acceptable as normal until age 3 years. Sustained ankle clonus or reflex asymmetries, however, remain pathological signs.

The specific newborn and infantile reflexes, however, are the province of pediatric neurology and provide information on normal neurological development as well as pathological changes. Their age of appearance and disappearance are shown in Table 2. The Moro reflex is the startle response, and is provoked by gently dropping the head in the supine infant held supported with the neck slightly flexed. There should be symmetrical upper extremity abduction, extension, and circumduction, and lower extremity extensive and then flexion. It is normally present at birth and involutes by 16-20 weeks. It is elicitable between 28 and 32 weeks in the premature, and serves as a clinical sign of neonatal encephalopathy when decreased. It is also a clue to a brachial plexus lesion, clavicle fracture, or hemiparesis when asymmetrical. The response should recede in time and should not be present beyond five months post-term.

The Galant is the truncal incurvation response. This is present between birth and three months of age. It appears by 24 weeks in the premature. The intact response is incurvation of the trunk ipsilateral to the side of stimulating the flank. The asymmetric tonic neck reflex (ATNR) describes the fencer's posture with ipsilateral arm extension and contralateral flexion upon rotation of the head to either side. A strong and long-lasting ATNR suggests pathology at any age, and the response is normally gone by six months. The sucking and rooting reflexes are present at birth. Sucking appears by 28 weeks and rooting, even more primitive, at 24 weeks of gestation.

A decrease in these responses suggests an encephalopathy. These responses disappear by the fourth month.

The palmar and plantar grasp responses are also present from birth. There is flexion of fingers and toes in response to gentle pressure to the palm and foot, respectively. The responses are clearly evoked in 32 week prematures. The so-called righting reflexes, present in the first four months, are responses by the infant to various movements and are part of the normal maturational response of developing protective responses. In the crossed leg extension reflex, the infant withdraws and flexes the stimulated leg and extends the other, as if pulling away from a threat and exerting antigravity resistance simultaneously. The neck-righting elicits an ipsiversive turning movement of the shoulders and trunk when the infant's head is turned. Similarly, there is a vertical righting response where the infant's lower extremities extend, followed by the trunk and head, when the infant is held flexed over the examiner's arm and the feet are stimulated.

In the parachute response, the infant extends the upper extremities toward the table as the examiner, supporting the prone infant, motions the head toward the examination table. This protective mechanism appears between 6 and 9 months of age and should be well-developed between 12 and 24

Table 2

Common Developmental Reflexes

Reflex	Age of Appearance	Age of Disappearance (post-term)
Moro	28-32 weeks GA	5-6 months
Rooting	24 weeks GA	4 months
Sucking	28 weeks GA	4 months
Grasp (palmar, plantar)	32 weeks GA	2 months
Galant (spinal incurvation)	24 weeks GA	3 months
ATNR	Birth	3-6 months
Crossed leg extensor	Birth	1 month
Placing	Birth	1-2 months
Stepping	10 days	1-2 months
Parachute	6-9 months	Persists
Landau (extends when supported prone)	10 months	24 months

GA = gestational age; ATNR = asymmetric tonic neck reflex

months. The Landau reflex, present by 10 months of age, evokes extension of the neck, trunk, and lower extremities when the infant is suspended by the examiner's hand in the prone position. Passive head extension results in further extension, and head flexion leads to flexion of the trunk and hips. Focal deficits such as hemiplegia or paraplegia, or exaggerated rigidity, may be detected by anomalous responses.

The antecedents to walking can be observed in the newborn reflexes. The steppage reflex is obtained by placing the infant's feet on a surface and alternating the shoulders forward to simulate walking. A heel take-off is reached by 40 weeks gestational age. In the placing reflex, the dorsum of the infant's foot is brought to the undersurface of a tabletop. The infant will raise the foot, placing it on the table in a stepping pattern. This reflex appears about ten days of age and its absence may indicate the presence of neurological deficits that are otherwise undetected by the examination.

Sensory

The sensory exam is particularly difficult in the child. Only major deficits will be detected in infants. In the young child, reliability of response is always a question, and the rhythm of testing should be altered to avoid automated answers. The preschooler can generally attend to double simultaneous stimulation and answer reliably enough to detect extinction of the stimulus on one side. Higher cortical modalities such as graphesthesia and stereoagnosia can be tested by seven to eight years of age.

Cerebellar Function

The cerebellar exam is traditionally directed toward this structure's role as a modulator of oscillatory activity, although functions related to frontal and striatal neural networks have greatly expanded our view of the cerebellum in clinical neurology. In the young child, offering toys will allow for evocation of cerebellar tremor or dysmetria. Asking the child to place a marble between the examiner's two fingers screens for incoordination. Other helpful maneuvers are having the child press the button on a tape measure, finger-to-nose testing, rapid alternating movements such as alternatively tapping the palm and dorsum of the hand on the thigh, or apposing the fingers to thumb as if playing the piano. The hands should be tested individually as the examiner assesses for the dysrhythmia of cerebellar disease, or the presence of extrapyramidal or adventitious movements suggestive of basal ganglia disease or dyspraxia. Excessive mirroring movements in the opposite limb represent so-called soft or subtle neurological signs, although extraneous movements may be normal until seven to eight years of age.

Gait is a screen of cerebellar function that signifies neurologic disease at all levels. Heel and toe gait can be reliably observed by four to five years of age. Impaired dorsiflexion as manifest by absent heel gait suggests a peripheral neuropathy. The waddling gait of pelvic girdle weakness is seen in muscle disorders, and coupled with oversized calves in a young boy suggests Duchenne muscular dystrophy. Excessive toe gait points to spasticity; absent toe gait may represent a lumbosacral spine lesion involving the conus medullaris or cauda equina. Impaired tandem gait implies problems with cerebellar pathways. A circumductive gait, coupled with decreased arm swing, suggests hemiparesis corresponding to a lesion of the contralateral cerebral hemisphere.

6. Conclusion

The history remains the cornerstone of the pediatric neurological examination. The chief complaint, more diplomatically referred to as the chief concern, is often best left in the family's own words. The history is best taken as a guided story provided by the family. The examination includes the general features, with emphasis on dysmorphic or cutaneous stigmata of a neurological disorder, and for the involvement of systemic organs. It is critical to account for the process of normal development, and its wide range of normalcy, during the enactment and interpretation of the pediatric neurological examination. As with the rest of clinical localization in the nervous system, the significance of the findings on examination depends upon the company that they keep. Are there supportive features in the history and examination to substantiate the finding under consideration? Are there related deficits in that developmental sphere to assign the presence of a neurological deficit? Are there deficits in other neurodevelopmental spheres to suggest the pattern of a syndrome? The examiner must continuously ask and reflect on these questions during the clinical evaluation. The neurological examination must emphasize simple observation and playful interaction with the child. While a successful examination requires both cleverness and originality, recruiting the child into challenging games invariably leads to an increased level of cooperation and rewarding physician-patient interaction.

7. References

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   This legendary manual is a must for pediatric residents and can ably assist the neurology house officer with practical synopses of all organ systems, including child neurology and development. There is an easily accessed formulary specifying pediatric (mg/kg) dosages.

2. Jones KL. *Smith's Recognizable Patterns of Human Malformations*. 5th ed. Philadelphia, Pa: WB Saunders; 1997.

   The bible of dysmorphism. This features a fact-filled page and accompanying set of photographs on the recognizable patterns of human malformation, as well as normative data for multiple anthropometric measurements.

3. DO Wiebers, AJ Dale, E Kokmen, JW Swanson, eds. *Mayo Clinic Examinations in Neurology*. 7th ed. St. Louis, Mo: Mosby; 1998.

   Both comprehensive and concise, this monograph covers the clinical neurologic examination and ancillary diagnostic procedures. A rich discussion of speech and language development and praxis is included in the chapter on the examination of infants and children.

4. Volpe JJ. *Neurology of the Newborn*. 4th ed. Philadelphia, Pa: WB Saunders; 2001.

   The classic for neonatal neurology. This includes authoritative sections on the orderly process of fetal brain development and congenital malformations, as well as hypoxic-ischemic encephalopathy and other neurologic problems of the newborn.

8. Questions

1. The findings of hepatosplenomegaly, petechiae, and microcephaly at birth are most consistent with:

   A. Perinatal hypoxic-ischemic encephalopathy

   B. Fetal alcohol syndrome

   C. Craniosynostosis

   D. TORCH infection

   E. Neonatal meningitis

2. Adrenoleukodystrophy is likely to present with which of the following patterns:

   A. Pigmentary retinopathy with seizures

   B. Cognitive decline

   C. Spasticity, ataxia, and visual loss

   D. Visual hallucinations

   E. Myoclonic epilepsy

3. In tuberous sclerosis, the initial cutaneous manifestation is likely to be:

   A. Adenoma sebaceum of the face

   B. Hypopigmented maculae of the skin

   C. Shagreen patch over the lumbosacral region

   D. Fibromata of the periungual nailbeds

   E. Pits of dental enamel

4. The hypotonic infant with hyperalert appearance is likely to have:

   A. Miller-Dieker syndrome

   B. Hyperekplexia

   C. Walker-Warburg syndrome

   D. Neonatal myasthenia

   E. Werdnig-Hoffman syndrome

5. Which of the following signs indicate an abnormality in a term newborn?

   A. Sustained ankle clonus

   B. Extensor plantar response

   C. Crossed adductor response

   D. Absent triceps reflex

   E. Closed posterior fontanelle

6. In the Landau reflex, there is extension of the neck, trunk, and lower extremities as the infant is held prone in the examiner's hand. This reflex normally appears by:

   A. Birth

   B. 1 month

   C. 6 months

   D. 10 months

   E. 12 months

7. The Moro or "startle response" is normally present during which of the following time intervals:

   A. Perinatal period

   B. Birth to 1 week

   C. Birth to 1 month

   D. Birth to 5 months

   E. 1 week to 6 months

8. The head lag that appears as a child is brought from the supine to seated position should resolve by age:

   A. 2 months

   B. 4 months

   C. 6 months

   D. 8 months

   E. 10 months

9. A healthy infant girl is able to pull herself up to stand and wave bye-bye. Her FOC measures 45 cm. What level of verbal language should be anticipated?

   A. Cooing (long musical vowel sounds)

   B. Reduplicated babbling (da-da-da)

   C. Says mama and dada nonspecifically

   D. Uses two words with meaning

   E. Combines two words

10. By age three years, a child is expected to:

    A. Copy a triangle

    B. Hop on one foot

    C. Distinguish left from right

    D. Distinguish morning from afternoon

    E. Pedal a tricycle

Answers

1. D.
The systemic findings of hepatosplenomegaly and petechiae in a newborn, associated with microcephaly, strongly suggest the presence of an in-utero viral infection. The latter represents a group of viral infections summarized with the acronym TORCH, ie, toxoplasmosis, syphilis (O is for "other"), rubella, cytomegalovirus (CMV), Herpes simplex, and HIV. There are some classic distinguishing features, such as the patterns of intracranial calcifications. The calcifications in CMV are typically periventricular, and in toxoplasmosis are intraparenchymal. In perinatal hypoxic-ischemic encephalopathy, dysfunction of other organ systems, eg, hepatic and renal, should be present as well. Microcephaly may be found in children with a history of perinatal hypoxic-ischemic encephalopathy after some time, but not at birth.

2. C.
Adrenoleukodystrophy (ALD), also known as Lorenzo's disease (note the movie, *Lorenzo's Oil*), is a peroxisomal disease presenting with adrenal insufficiency and progressive dysmyelination which proceeds in a posterior to anterior distribution. Leukodystrophies such as ALD (as well as metachromatic leukodystrophy or MLD, or Krabbe's disease) will present with long tract signs such as spasticity and ataxia, as well as visual loss due to optic atrophy. In contrast, gray matter degen-

erative diseases will present with primary neuronal symptoms, such as cognitive decline and seizures. Option A, pigmentary retinopathy with seizures, would be classic for neuronal ceroid lipofuscinosis (NCL), of which Batten's disease is the type affecting school age children. Option E, myoclonic epilepsy, would be more representative of a gray matter than white matter disorder.

3. B.
The cutaneous findings of tuberous sclerosis complex include all of the listed items, although the earliest to appear are the hypopigmented maculae of the skin traditionally known as ash leaf spots. This descriptive term is somewhat amorphous, as ash leaves do not have a distinct shape. In any case, "hypopigmented maculae" is more descriptive and less confusing. Their detection is aided by the use of the fluorescent Wood's lamp in a dark room, particularly in individuals with light complexions. This is worthwhile in screening first degree relatives of patients, as this multisystem phakomatosis has wide phenotypic variation and asymptomatic parents are sometimes detected during the evaluation of their affected child.

4. E.
Hypotonia in an infant can be due to either an upper or lower motor neuron lesion. The association of profound hypotonia in an unusually alert appearing infant is classic for spinal muscular atrophy. Type I, with onset within the first six months of life, is known eponymically as Werdnig-Hoffman syndrome. These patients will have absent reflexes. The intermediate form, Type II, has onset after six months and features tongue fasciculations. The adolescent form is known as Kugelberg-Welander syndrome.

Miller-Dieker and Walker-Warburg syndromes are genetic forms of lissencephaly, a congenital brain anomaly of disordered neuronal migration leading to a smooth cerebral cortex. Hyperekplexia is a startle syndrome related to a mutation of the glycine inhibitory receptor. Neonatal myasthenia is a transient condition related to passive transfer of acetycholine receptor antibody from a myasthenic mother. This is in contrast to congenital myasthenia, which refers to a group of genetic disorders leading to abnormal formation of the neuromuscular junction.

5. A.
Sustained clonus would represent an abnormal neurological finding in a term newborn. An extensor plantar response is expected up to one year of age and acceptable up to three years of age. The triceps reflex may not be seen until six months of age in normal infants. An open posterior fontanelle at birth would raise concern about congenital hypothyroidism or increased intracranial pressure.

6. D.
The newborn and infant reflexes represent developmental patterns that presage later maturation. These appear and extinguish within normal time ranges, and late appearance or disappearance suggest abnormal development. The Landau reflex, manifest by extension as the infant is held prone, appears by ten months and disappears by two years of age.

7. D.
The newborn's startle response, known as the Moro, is present at birth. Absence may indicate a neurologically depressed infant; asymmetry suggests a hemiparesis, brachial plexopathy, or clavicle fracture. The response is typically gone by five or six months of age.

8. B.
By four months of age, an infant should have enough head support to maintain the cranium in line with the trunk while raised by the hands from supine to sitting. Persistent head lag suggests hypotonia, of either central or peripheral localization.

9. C.
This vignette depicts a normal nine month old. The cooing of long vowel sounds is heard at about three months of age. Reduplicated babbling, ie, ba-ba-ba, da-da-da, is present about six months of age, and combined syllables (nonreduplicated babbling) by eight months. Discriminate use of dada or mama is typical at 10 months. By one year, a child is expected to use two words with meaning (other than dada/mama). Two-word combinations typically appear at about 18 months.

10. E.
Three-year-olds ride three-wheel bikes! Of the "Gesell figures," a 15-month-old imitates a scribble; an 18-month-old scribbles spontaneously; a 2-year-

old imitates a stroke; a 2-1/2-year-old differentiates a horizontal versus vertical line; a 3-year-old copies a circle; a 4-year-old a square; and a 5-year-old a triangle. A diamond is mastered at ages 6-7 years; cylinder while age 9; and a cube at age 11. A 4-year-old learns to hop. Left versus right and morning versus afternoon distinction are the province of the six-year-old.