Gillette Children's Specialty Healthcare

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Red Flags and Rules of Thumb:

Sorting Out Developmental Delay

by Raymond Tervo, M.D.

Each meeting between a physician and a child is an opportunity to assess developmental and behavioral issues. Timely detection of any potential problems is important, because brain development is most malleable early in life.

Red flags and rules of thumb can simplify the task of identifying delayed development. Red flags signify warnings. Rules of thumb are principles with broad applications that are not intended to relate to every situation. The rules are easily learned and easily applied.

The purpose of this article is to outline red flags and to describe some clinically relevant rules of thumb about development in children.

The Role of Parents in Assessing Development

A thorough history is essential to understanding developmental concerns.[1] When a parent is worried, there is a good chance that something needs attention.[2,3] Parents' concerns are particularly likely to be accurate in the areas of speech and language skills, cognitive performance, and fine motor skills.[2]

William Osler once said, "Listen to the patient; he is trying to tell you the diagnosis." One rule of thumb is: never ignore a parent's concerns. Always suspect delays — and, when they are observed, refer early. Developmental screening using parental reports or other instruments can be more accurate than clinical judgments.[4,5]

Clinicians should closely monitor children and families with risk factors. Promptly refer a child who has an established diagnosis associated with developmental delay (such as Down syndrome or autism). There are comprehensive lists of organic and psycho-social risk factors, but among the most worrisome are a very low birth weight, a clear case of asphyxia, and neonatal complications. Remember: the lower the birth weight, the greater the chance of morbidity.[6]

Red Flags in Development

When assessing a child's development, it is essential to pay attention to serious neurologic syndromes or other conditions that raise important red flags.

Serious neurologic syndromes include unexplained focal neurologic features, developmental regression or marked stagnation, seizures with developmental implications, concerns with head circumference, and unusual skin and facial features.[7]

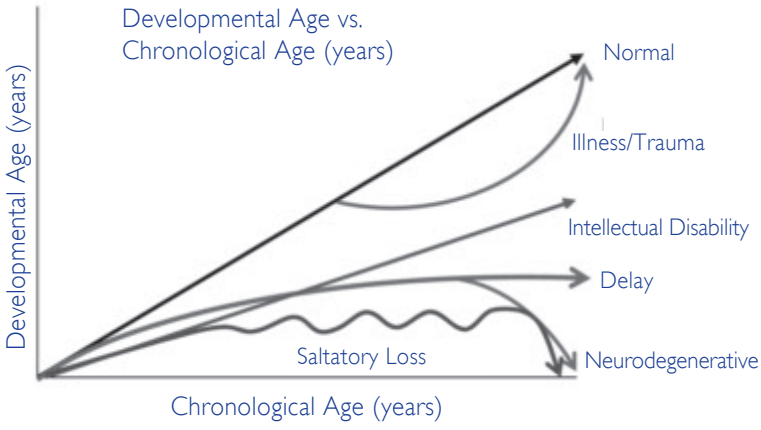
Focal Lesions

A focal lesion must be considered in a seizure disorder. Focal lesions are likely in hemiparesis, ataxia, and blurred or double vision. As in children with autism and attention deficit hyperactivity disorder, children with global developmental delay virtually never have focal lesions.

Regression and Stagnation

When there is regression or marked stagnation, delayed development is a concern (Figure 1). Most presentations of global developmental delay result from a previous injury or disease and remain stable over the years.

Figure 1: Developmental Trajectories



Static abilities are rarely normal and are therefore a red flag. If children's abilities do not improve with time, those children might be regressing. It is important to recognize that developmental delay or regressing skills can be caused by raised intracranial pressure secondary to a tumor,

Continued on Page 2

hydrocephalus or other non-neurodegenerative entities.[8] If there is regression, clinicians must consider whether the case involves true regression or the evolution of signs in a static encephalopathy, such as autistic regression.[8]

When a child loses developmental milestones, watch for signs and symptoms of meningitis. Unexplained persistent loss of neurologic function is always worrisome. It might include such presentations as a child losing the ability to walk or an unexplained change in mental status (such as occurs in a school-aged child with adrenoleukodystrophy who fails to respond to psychostimulants for attention problems).

There are other presentations that raise important red flags. Severe delays can point to a condition such as Rett syndrome. Absent reflexes or a positive Gower's sign can be signs of spinal muscular atrophy or Duchenne muscular dystrophy. Midline lumbosacral abnormalities, such as sacral dimples, might indicate occult spinal dysraphism; other spinal or orthopaedic abnormalities can suggest a neuromuscular cause of delay. A dysmorphologic evaluation might reveal a syndrome.[9]

Seizures

Psychomotor regression is often a feature of severe epilepsies, such as Lennox-Gastaut syndrome or infantile spasms. Infantile spasms are a variety of generalized, myoclonic seizures marked by brief motor spasms of the head and extremities. The worry is that there might be an underlying neurologic disease, such as tuberous sclerosis, cerebral dysgenesis, and hypoxic-ischemic encephalopathy.

Head Circumference

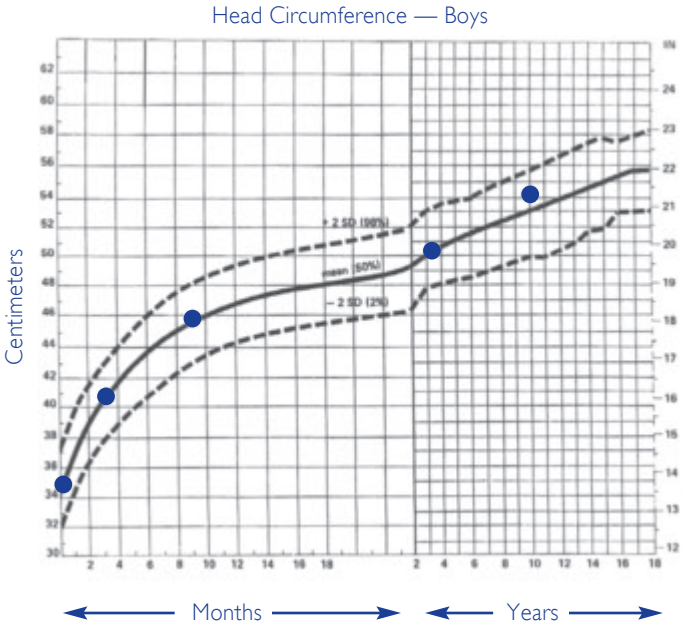
One of the most important measures on physical examination is head circumference (HC), because HC correlates with brain size and growth of the infant.[10] A child with a small HC is microcephalic; a child with a large HC is macrocephalic. Macrocephaly, delay and regression might suggest Canavan/Alexander, Krabbe or metachromatic leuko-dystophy. Microcephaly with delay and regression might be a clue to Rett syndrome or infantile neuronal lipofuscinoses.

There is cause for concern when a newborn's HC is substantially less than his or her chest circumference. A rule of thumb for HC is the "three and nine" rule (Figure 2): a newborn has a HC of 35 centimeters (cm); a 3-month-old has a HC of 40 cm; a 9-month-old has a HC of 45 cm; a 3-year-old has a HC of 50 cm; and a 9-year-old has a HC of 55 cm.[11] Record whether the HC falls above or below the second percentile and whether it crosses percentiles.

Skin and Facial Features

Other features on physical exam ought to give cause for concern. Main syndromes with typical facial features include

Figure 2: "Three and Nine Rule" for Head Circumference



fetal alcohol and Down syndrome. Skin findings — including signs of a neurocutaneous disorder, such as hypo- or hyperpigmentation — should be noted. An examination of the skin for café au lait macules or ash-leaf spots enhanced with a Wood's lamp examination can help diagnose neuro-fibromatosis or tuberous sclerosis. The fundi might show a cherry-red spot with hepatomegaly and alert clinicians to the presence of a lysosomal disorder, such as Tay-Sachs.

Measuring Development

A meticulous developmental assessment is warranted to evaluate development. Developmental delay occurs when there is a "substantial" delay in one developmental domain or more. "Substantial" implies functional deficits that are equal to or greater than two standard deviations below the mean for chronological age (after correction for prematurity).[12]

Calculate the child's developmental quotient (i.e., the ratio of developmental age to chronological age). A useful rule of thumb is: if the ratio is greater than 85, for the moment one may reassure the family; ratios of 75 to 85 deserve close monitoring; children with ratios of less than 70 ought to be referred. Note that developmental quotients are reported as ranges. Any attempt to be more accurate — for example, to differentiate a 71 from a 70 — can only lead to error.

Another rule of thumb is that delays in one developmental domain can impair development in another and can impair

the clinician's ability to evaluate those skills. Development occurs as overlapping waves, so that a child may appear to progress in one area of development but then forego interest in that area while working on another developmental domain. A rule of thumb is that, in the first year of life, a child who is two months delayed beyond the expected mean for that age ought to be evaluated in more detail or referred. From 1 to 2 years of age, the window is four months.[13] Global developmental delay occurs when there are substantial delays in two or more developmental domains.[14] Each area of development should be assessed carefully, because one area of obvious delay — such as gross motor — can effect more subtle deficits in other areas.[8]

Language Delays

It is important for clinicians to keep typical language milestones in mind. Red flags of language development necessitating referral include:

- No babbling, no pointing, no gestures by 12 months of age
- No single words by 16 months of age
- No nonecholalic two-word sentences by 24 months of age
- A loss of language or social skills at any age[3, 15]

A rule of thumb for atypical verbal or auditory language milestones is that one ought to refer a child who, at 2 years of age, has a vocabulary of fewer than 50 words and creates no two-word sentences.[16]

In evaluating expressive language development, the rule of four is a particularly useful rule of thumb:

- At age 1, a child uses single words and is intelligible to about 25 percent of strangers.
- At age 2, a child uses two-word combinations and is intelligible to about 50 percent of strangers.
- By age 3, a child uses three-word sentences and is intelligible to about 75 percent of strangers.
- By age 4, a child uses four-word sentences and should be intelligible to all strangers.[17]

As a rule of thumb, language comprehension nearly always comes before language expression. When comprehension is much more advanced than expression and a child has difficulty executing oral movements but has intact auditory comprehension skills, however, consider childhood apraxia of speech.[15] On the other hand, "advanced" expressive skills and delayed receptive ones can be seen in autism.[3]

When reviewing language development, do not overlook hearing. Deaf babies gurgle and coo, and their early language development might appear to be normal. All newborns should be screened for hearing loss by 1 month of age and have any hearing loss diagnosed by at least 3 months of age. Amplification should begin within one month of diagnosis. Always refer to an audiologist if the child is delayed.

Language assessment is the most sensitive indicator of intellectual disability in preschoolers. Autism usually manifests as language delay. The key message is that, when there is worry about a child's language delay, assess hearing and then refer for a more detailed assessment. When deafness is suspected, only a complete hearing test is conclusive.

Motor Delays

The key importance of delayed motor development lies in alerting clinicians to signs of cerebral palsy, hypotonia or muscular dystrophy.

Physical and motor development occurs in a cephalocaudad direction (that is, downward from head to feet) and a proximodistal direction (that is, from a child's torso to the hands and feet). A traction response in early infancy is especially worrisome because it indicates hypotonia, which can be a sign of an underlying congenital myopathy.[12]

Early acquisition of motor skills almost always excludes an intellectual disability. Shuffling or hitching delays the onset of walking. Blind children need to be taught to walk. Children with benign congenital hypotonia walk late, as do children with Duchenne muscular dystrophy. Check creatine phosphokinase levels in all boys who are not walking by 18 months of age.

Generally, myopathies cause proximal weakness, and disorders of peripheral nerves lead to distal weakness. A child who has myelomeningocele with severe lower-limb involvement will be unable to walk without skilled orthopaedic intervention. Walking will be delayed in children who have conditions associated with excessive muscle tone, such as cerebral palsy. As a rule of thumb, children who have an intellectual disability but no cerebral palsy can be expected to walk; when motor delay is considerable, there is usually a cause.

The following red flags in motor development might be early signs of cerebral palsy. A child who rolls at younger than 3 months might be hypertonic, and opisthonic posturing might appear as an effort to roll. At 4 months, a child who lacks head control and who demonstrates scissoring (indicating adductor tightness) might be worrisome. At 6 months, the persistence of primitive reflexes, which should have integrated and disappeared by then, is a troublesome sign. At 7 months, the fact of W-sitting spasticity and hypotonia deserves a more careful look. A child who is not sitting well at 9 months or not pulling to a stand by 12 months is a child who needs a thorough evaluation.

Remember that cerebral palsy is the most common neuromotor disability in childhood. When there is no motor delay, there is no cerebral palsy.[18] When evaluating a child with

cerebral palsy, it is important to communicate the severity of the condition to parents. A useful rule of thumb for describing functional deficits is that a child with mild cerebral palsy is one who ambulates without assistance. A child with moderate cerebral palsy requires aids and appliances. A child with severe cerebral palsy is mobile only in a wheelchair. The average age of walking in the different forms of cerebral palsy is hard to gauge, because intelligence has a profound effect on motor development.

Visual Fine-Motor Skills
It is important to evaluate visual fine-motor skills. In the first three months of life, visual skills are crucial. All babies with apparent visual impairments should be seen by an ophthalmologist. Late smiling might suggest blindness, autism, or conditions such as Mobius syndrome or myotonic dystrophy. Hand dominance prior to 18 months of age is a red flag.

Some important milestones concerning visual skills include the presence of blink or papillary light reflex, which appears at around 31 weeks' gestation and should be present at birth. By 6 to 8 weeks of age, a child should be able to demonstrate fixation and have straight eye alignment. By 3 months, fixation should be established; a child should track and have conjugate eye movements. From 3 to 6 months of age, a child should reach for objects and show smooth tracking.[19]

It is often difficult to diagnose a mild strabismus, but it is of critical importance to do so. A persistent squint at any age is abnormal, and a new squint warrants urgent attention. Always be concerned about intracranial pressure. An asymmetric Moro reflex or, later, a failure to reach for objects might indicate a brachial plexus injury. The most common peripheral nerve injury is Erb's palsy caused by shoulder dystocia or a difficult delivery.

Delayed visual maturation can indicate a developmental disability. Dyspraxia is unlikely to be a key symptom, but it is prevalent in neurologic disorders and the symptoms depend on age. Dyspraxia, mild cerebral palsy and motor delays can disadvantage development in general.[8]

Cognitive Delays
There are significant milestones for cognitive development. By 9 to 10 months, a child should have established object permanence. By 18 months, a child should display joint attention. At about 18 months, protolinguistic pointing and symbolic play are noted.

When a child's speech is delayed, always worry about a cognitive disability. One rule of thumb when considering a young person who presents with school problems is to evaluate the patient's IQ-achievement discrepancy. A child is learning-disabled if achievement is two or more grades behind the grade level expected for IQ and age.[13]

Behavior Problems
A rule of thumb for social and behavior problems is that a child's social and emotional development correlates with language development. A child's behavioral problems warrant professional help if they are persistent, prolonged, and interfere with the ability to function at home or at school.

Summary

If there is the slightest doubt about a child's developmental assessment, that child must be seen again. Observing the rate of change in development is essential. In many instances, judgment about a child's development must be withheld until a reassessment occurs.

Developmental diagnosis is difficult. It is important to avoid overconfidence, making spot diagnoses, and failing to follow a child's developmental trajectory. Similarly, offering a long-term prognosis following an initial development assessment is likely to be fraught with error. An evaluation allows one to make a statement about a child's capacities and promise, but it is not possible to say what the child will do those qualities.

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Author's PROFILE

Raymond Tervo, M.D., is a neurodevelopmental pediatrician and medical director of Pediatrics at Gillette Children's Specialty Healthcare. He earned a medical degree at McMaster University in Hamilton, Ontario, Canada, and did his core pediatric residency training at the Hospital for Sick Children in Toronto. He also completed his subspecialty training in developmental pediatrics at McMaster, and he is board-certified in neurodevelopmental disabilities.

Dr. Tervo has served on the faculty of the University of Saskatchewan, the University of Toronto and the University of South Dakota. He is an associate professor in the department of Pediatrics at the University of Minnesota. His professional memberships include the Society of Developmental and Behavioral Pediatrics and the American Academy for Cerebral Palsy and Developmental Medicine.

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At the half-day course on Oct. 8, presenters will discuss research findings. In the full-day course on Oct. 9, attendees will participate in small, clinical forums with scientists. The forum discussion will focus on the implications of current research evidence for clinical applications.

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For information on course content, call John Belew at 651-726-2621 or 800-719-4040 (toll-free). Or e-mail him at jbelew@gillettechildrens.com. For registration information, call Mary Grimm at 651-578-5002 or 800-719-4040 (toll-free). Or e-mail her at mgrimm@gillettechildrens.com.

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