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EXPERT REVIEWS

Delayed visual maturation: a visual inattention problem

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Delayed visual maturation (DVM) is a term used to describe infants who do not exhibit the ability to fix or follow objects in the environment, but subsequently improves by the age of 6 months without treatment. Infants with these features were first described in the 1920s by Beauvieux, but today there remains little consensus as to the etiology of this phenomenon. This article reviews the existing literature on DVM and summarizes what is known about the classification, clinical characteristics and prognosis of this condition. In this article diagnosing DVM is discussed as well as a summary of other conditions to consider on the differential. Current thought on the etiology of this condition is explored. Ocular examination, electrophysiology tests, imaging results and central eye movements appear normal. These findings imply anterior and posterior sensory pathways as well as cortical motor pathways are intact in these patients. Given that these pathways are intact, a possible explanation for these symptoms includes temporary injury to the attention centers in the developing infant's brain. Further research is needed to better understand the causes of this condition as well as the long-term sequela.

Keywords: delayed visual maturation • developmental delay • temporary visual inattention

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An infant who presents with isolated delayed visual maturation (DVM) is cause for great anxiety for parents. The child appears blind, unable to focus attention, fixate or follow any part of the visual world. Ocular and neurologic examination of the child is completely normal and visual behavior resolves rapidly (sometimes overnight) by 4–6 months of age. Symptoms resolve without treatment and often, no cause for the initial presentation is found.

The pathophysiology of isolated DVM is unknown and represents a spectrum disease with several possible entities. Cases of DVM were first described in other terms by Beauvieux and referred to as 'temporary visual inattention' [1,2]. The author described an anomalous, gray appearance of the optic nerve in these cases and postulated that this was due to a problem with myelination. He called this phenomenon 'pseudo-atrophie optique' and 'dysgenesie myelinique' [2]. Doggart *et al.* also described DVM infants and implicated a delay in myelination of the optic nerve [3].

Illingworth first introduced the term 'delayed visual maturation' to describe the phenomenon [4]. He described two infants who initially took no notice of their surroundings, but gained signs of vision by 5–6 months of age. Both infants were not developmentally delayed, with the exception of one infant with delayed walking at the age of 2 years. Also of note, this infant had spasmus nutans (transient nystagmus) with slight residual signs at the end of follow-up.

True isolated DVM is a relatively rare phenomenon, but the prevalence is unknown. The cause is not well understood and there is no clear evidence that any primary visual system is delayed. Some authors believe that this entity represents a primary visual inattention disorder [5].

Classification

The term DVM is usually used to refer to isolated (type I) DVM in the clinical setting. For the purposes of this article, we will describe the clinical characteristics and classification of all types of DVM and then return to a discussion of isolated DVM.

Beauvieux first classified the entity now known as DVM into two groups. The first group had DVM as the only anomaly with recovery rapid and complete. The second group included ocular problems such as strabismus, high refractive error as well as mental retardation, resulting in slower and often incomplete recovery [2]. Next, DVM was classified into three groups: group I: simple visual developmental delay; group II: visual developmental delay associated with mental retardation and group III: visual developmental delay associated with ocular abnormalities [6].

Group I was expanded by Fielder *et al.* to include two subgroups: group A: DVM as the sole abnormality and group B: DVM with 'concurrent systemic illness or history of perinatal problems' [7]. The most common perinatal abnormalities presented in group IB included birth asphyxia and 'jitters/fits'. In addition, group IA was further divided into IAi: infants with poor vision on presentation to the doctor and IAii: infants that improved by the time they presented to the doctor and were diagnosed by history [7].

The classification system was again modified to its current form to separate group III into type III, which includes congenital nystagmus and albinism and type IV, which includes other severe ocular disorders [8]. See **Box 1** for summary of classification.

Clinical characteristics: type I DVM

Type I DVM is characterized by infants that do not have the ability to fix or follow objects in the environment. They are usually brought in for medical care when their symptoms do not resolve by 2–4 months, although no consensus exists on when diagnosis should be made. The diagnosis is confirmed after symptoms resolve and other visual disorders are excluded. DVM is characterized by a rapid resolution of symptoms by age 6 months.

Infants with DVM have no detectable ocular or neurologic abnormalities to explain their symptoms. Strabismus is often present during the phase of visual impairment, most commonly divergent and less frequently convergent [9]. Pupils are normal and ocular examination is usually normal. However, as described by several authors, the optic disc has been noted to be gray compared with usual [2,10].

Electrophysiology results

Several studies have shown normal electroretinogram (ERG) results in patients with DVM, including two studies that had

eight and nine patients respectively [11,12]. By contrast, there is little consensus regarding visual evoked potential (VEP) results for this patient population. An early study of four children with normal ERGs showed initially impaired visual evoked responses, but normal responses by the age of 4 months [13]. Harel *et al.* described three infants with DVM whose VEPs were of prolonged latency, but repeated VEPs showed improvement in latency and waveforms after the age of 6 months [14].

Other authors such as Lambert *et al.* demonstrated no significant difference in the latency or amplitude in VEPs when compared with age-matched controls [12]. Another study showed abnormal flashed-evoked potentials in DVM infants that improved with resolution of symptoms [15]. More recently, a case series of two DVM infants described normal thresholds for vernier and grating acuity [16]. It is unclear whether DVM infants have abnormal VEPs, normal VEPs compared with age-matched controls or perhaps the entity may present with a spectrum of VEP results. Whatever the case, the appearance of an abnormal VEP before age 4 months is unlikely to dictate the infant's prognosis [13].

In another study, grating acuity was estimated using Teller Acuity Cards and showed normal results in infants with DVM [17]. However, a previous study showed 26 infants with type I–IV DVM with abnormal grating acuity [9].

Imaging results

Neuroimaging studies of patients with DVM have thus far indicated no significant difference between these patients and controls. An MRI study investigating 14 infants with type I DVM, evaluated by T1-weighted evaluation of optic nerves and chiasm showed no difference between these infants and controls [18]. However, a delayed myelination pattern was found in three cases. Mercuri *et al.* showed one infant with DVM type Ib with diffuse white matter changes and bilateral lentiform nucleus lesions [19].

Central eye movement characteristics

The defining characteristic of DVM is the inability to fix and follow. However, other central eye movement systems appear to be intact. Vestibular-ocular response (VOR) is observed in infants with DVM. Two out of eight infants in one study showed a completely normal VOR, while the rest only showed a lack of fast phase [11]. This seems to be consistent the normal pattern in preterm infants as displayed in other studies [20]. Only one out of nine infants in one study demonstrated a lack of VOR, as well as optokinetic nystagmus (OKN) that persisted [12].

Saccadic movements are not present in the absence of fixing and following eye movements, but as this function returns, saccadic movements follow. In a study of infants with DVM, Harris *et al.* showed that all six had normal binocular full-field OKN, despite the inability to fix and follow [21]. This suggests that the oculomotor centers for generating smooth eye movements and brainstem saccadic function are intact [21]. However, immature monocular OKN is found in infants with DVM compared with controls and some authors suggest a cortical deficiency as one possible mechanism [22].

Box 1. Classification of delayed visual maturation.

Type I
- DVM as only abnormality
- Normal development, perinatal period
- Perinatal problems or systemic illness

Type II
- DVM with residual neurologic abnormalities

Type III
- DVM with congenital nystagmus and albinism

Type IV
- DVM with other severe ocular abnormalities

DVM: Delayed visual maturation.
Data taken from [6–8].

Transient nystagmus has been reported in the literature in infants with DVM, although this seems to represent the minority of cases [23]. A study of 16 infants with DVM showed three with transient nystagmus [24]. Conversely, in a study of 11 infants with transient nystagmus, only two also had DVM, demonstrating that DVM possibly explains a minority of transient nystagmus cases.

Associated developmental problems

Delayed visual maturation represents a spectrum of disease and there are multiple instances where infants with presumed isolated DVM also had other developmental delays. As previously mentioned, one of the first cases Illingworth described with DVM had a transient delay in motor functioning that resolved [4]. Of the 16 children described by Cole *et al.*, two had delays in speech development [24]. Hoyt *et al.* described eight children with DVM, with seven described as having general "delays in motor development" [11]. Six out of these eight children were premature or small for gestational age. Of the nine DVM infants in the Lambert *et al.* study, four were described as having mild delay and one with severe delay [12].

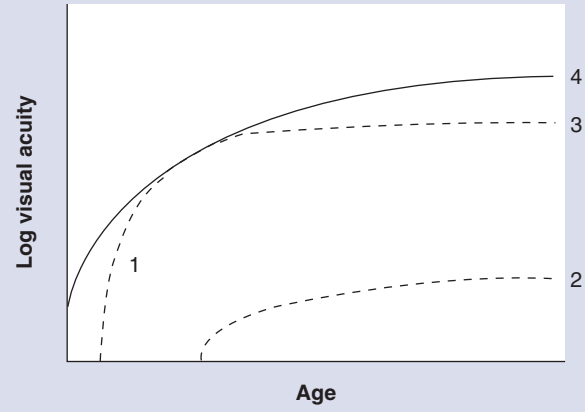

Figure 1. Schematic representation of acuity development in delayed visual maturation. In delayed visual maturation (DVM) type I, visual acuity improves rapidly and returns to normal (line 1 connecting to line 4). In DVM type III, visual acuity also improves rapidly but returns to near-normal (line 1 connecting to line 3). In DVM types II and IV visual acuity is late to improve and remains impaired (line 2).
Reproduced with permission from [42].

The diagnosis of DVM type I implies that infants with this disorder will regain visual function and have no future problems, but these early case reports indicate other consequences. A retrospective study by Hoyt of children diagnosed with isolated delayed visual maturation from 1981 to 2001 revealed a high level of neurodevelopmental and educational problems [5]. This study included 98 infants with at least 3 years of follow-up. A total of 93 of those patients had excellent visual acuity (20/20), but 22 had learning disabilities and 11 had a diagnosis of attention deficit disorder [5]. This suggests that this disease does not have a completely benign outcome.

It may be possible that some infants originally categorized as having isolated DVM actually have other associated delays or even DVM type II. Longer follow-up often reveals this link.

Type II DVM

Type II DVM is characterized as infants with attention and fixation deficit that additionally have neurological and or learning abnormalities. Resolution of visual symptoms is slower and often incomplete compared with DVM type I [9]. Many infants in this classification have mental retardation associated with seizure disorder or infantile spasm related to 'asphyxia, hypoglycemia, hypocalcemia, tuberous sclerosis, Aicardi's syndrome and so on [25]. Visual symptoms often improve when seizures are treated. Other causes of mental retardation not associated with seizures less often exhibit DVM [25].

We suggest that infants in this category often exhibit signs of DVM owing to damage (often hypoxic) to the extrageniculostriate visual system that predominates in early infants [22]. Symptoms may subsequently resolve when the geniculostriate system develops.

Full-field OKN are probably impaired in this population and neuroimaging is probably abnormal. Nystagmus may be present and could be due to another neurologic cause [22].

Type III DVM

Infants in this category have associated congenital nystagmus and albinism. Their vision starts to improve later than infants with DVM type I and improve to low-normal levels [9]. Often, nystagmus is absent during the period of DVM and can appear uniplanar with slow large oscillations at the time of visual recovery [26]. Other presentations of DVM type III include chaotic eye movements with poor fixation and large-amplitude pendular nystagmus [26].

Type IV DVM

Severe ocular disorders included in the DVM type IV classification include: retinal dystrophies, optic nerve hypoplasia and macular coloboma [22].

Prognosis & resolution of symptoms

Resolution of vision appears to take progressively longer when comparing higher group I with group II and III. Tresidder *et al.* demonstrated varying rates of return to normal vision based on the classification of the patient [9]. Under the classification system used by Tresidder *et al.*, visual improvement commenced the following week (mean): group IA – 15 weeks, group IB – 15.7 weeks, group II – 45.7 weeks and group III – 21 weeks. This study also demonstrated that all infants in group I regained normal vision, none in group II regained normal acuity and all infants in group III attained low-normal visual acuity.

Fielder *et al.* described similar time of improvement for type I, II and III DVM and also noted a wide range of improvement in type IV infants (5–46 months of age) [8]. A study of 11 children with type IV DVM (five Leber's congenital amaurosis, four optic nerve hypoplasia and two macular colobomata) showed three that remained blind (two Leber's congenital amaurosis and one optic nerve hypoplasia) [27]. No clinical characteristics were identified as predictive.

As shown in Figure 1, DVM type I (lines 1 and 4) and type III (lines 1 and 3) show quick improvement of symptoms. Whereas type I improves to normal, type III does not exhibit completely normal visual acuity later in childhood. Type II and IV show variable improvement depending on the severity of the underlying dysfunction. In general, signs of improvement start late and may reach a significantly impaired, but functional level (line 2). However, there may be further deterioration in later life given the nature of the disorder.

Isolated DVM-associated conditions

Delayed visual maturation is associated with other conditions such as prematurity, perinatal problems and possibly subtle structural brain damage. It is possible that in isolated DVM, these insults are subtle enough to only cause visual symptoms and not present with neurologic abnormalities.

In addition to perinatal problems such as hypoxia or periventricular hemorrhage, there may be other maternal factors that could contribute to DVM. In a study of 13 cocaine-exposed infants four had isolated DVM, suggesting a role of dopamine in the development of visual attention [28].

Another study of Karen refugees in Thailand noted abnormality in tracking and focusing compared with British control infants [29]. These abnormalities corrected by the age of 4–5 months and low levels of micronutrients such as vitamin A, vitamin C, folate and thiamine were noted in this population. Although no cause for early visual deficiency in this population was found, these authors postulated that early differences are possibly the result of nutritional deficits during critical periods of gestation [29].

Isolated DVM diagnostic approach

Evaluation of an infant lacking visual focus and attention requires careful ophthalmologic examination including anterior segment, lens and fundus. DVM is a diagnosis of exclusion and requires ruling out other possible causes. The initial examination should therefore include refraction. A case report of two infants with visual inattention showed immediate recovery after optical correction [30]. Spectacle correction was performed on the two infants at 5 and 4 months and they had -4 diopters (D) oculus dexter (right eye; OD), -5 D oculus sinister (left eye; OS) and -9 D oculus uterque (both eyes; OU) myopic error. In addition to careful

Table 1. Differential diagnosis of delayed visual maturation.

DVM and differential diagnosis: clinical characteristics	Eye examination	Pupillary reflex	Improves on refraction	Saccades	Seizures present	Neuroimaging	VEP	OKN/VOR	ERG
Delayed visual maturation	Normal, occasional gray discs	Normal		Intact	Possible, not likely	Normal	Normal	Normal	Normal
Cortical visual impairment	Usually normal	Usually normal		Intact	Likely present	Likely abnormal	Some abnormality	Some impairment	Normal
Ocular motor apraxia	Usually normal	Usually normal		No normal voluntary horizontal saccades Vertical saccades intact	Usually not		Usually normal	Impaired OKN, no fast-phase VOR	Usually normal
Saccadic palsy	Usually normal	Usually normal		No ability to make saccades	Usually not		Usually normal	No fast-phase VOR	Usually normal
Hereditary retinal dystrophy (including Leber's congenital amaurosis)	Normal early, can develop subretinal black pigment later	Some abnormality		Intact	Usually not				Undetectable or severly impaired
Global developmental delay	Could be normal	Usually normal			Usually not				
Epilepsy	Could be normal	Usually normal			Characteristic				
Optic nerve hypoplasia	Pale or gray disc, smaller than normal	Usually normal			Usually not				
Cocaine exposure	Could be normal	Usually normal			Usually not				
Autism	Could be normal	Usually normal			Usually not	Variable			Normal
High refractive error	Could be normal	Usually normal	Characteristic		Usually not	Normal			Normal

DVM: Delayed visual maturation; ERG: Electroretinogram; OKN: Optokinetic nystagmus; VEP: Visual evoked potential; VOR: Vestibulo-ocular reflex.
Data taken from [5,22].

ophthalmologic examination, a careful perinatal history must be taken, including any possible hypoxic events or any maternal substance abuse during pregnancy.

Early in the evaluation, it should be noted whether spontaneous nystagmus is present. No nystagmus indicates a normal anterior visual pathway. If nystagmus is present, however, careful examination of the eye as well as neuroimaging should aid in the diagnosis. The next diagnostic algorithm indicated by some authors indicates testing VOR and watching for saccadic eye movements [5]. If the fast motion movement is absent, then this suggests a deficiency in the saccadic mechanism. Global saccadic palsy or ocular motor apraxia should be considered [5]. If VOR is normal, then cortical visual impairment versus DVM should be considered and neuroimaging is necessary to resolve the difference.

Although this diagnostic algorithm is useful, it is also worthy to note that exceptions can occur along the way. For example, cases of transient nystagmus are noted in some infants with DVM [31]. In addition, another study showed six out of eight DVM infants with a lack of fast-phase VOR, which could be normal depending on the level of prematurity [11]. Other testing that could help with the diagnosis include ERG (a normal result rules out retinal dystrophy), OKN and Teller acuity.

One of the hallmarks of DVM is that the child's visual symptoms improve by 5–6 months of age. In the absence of any abnormality by eye examination, ERG, VOR, OKN and no history of perinatal complication, it is prudent to continue with neuroimaging if symptoms do not show signs of resolution by 5–6 months of age. MRI characterization of the lesion establishes the diagnosis and prognosis for the patient [32].

Approach to parents

On initial presentation, parents need to understand that evaluation of children with DVM involves a multidisciplinary approach between the ophthalmologist and pediatrician. If there are no other abnormalities noted, (isolated DVM), then the parents need to be reassured that prognosis is good for their child's visual acuity. Symptoms should resolve by 6 months of age with no increased risk of visual abnormalities in the future. However, not much is known about the long-term developmental sequelae in these patients and regular check-ups with a pediatrician are recommended.

Box 2. Differential diagnosis for isolated delayed visual maturation†.

- Cortical visual impairment
- Ocular motor apraxia
- Saccadic palsy
- Hereditary retinal dystrophy (including Leber's congenital amaurosis)
- Global developmental delay
- Epilepsy
- Optic nerve hypoplasia
- Cocaine exposure
- Autism
- High refractive error

†See Table 1.
Data taken from [5,22].

Isolated DVM differential diagnosis

The differential diagnosis (see Box 2) for an infant that is apparently blind is initially broad. However, after appropriate ophthalmologic evaluation and testing, this list can be narrowed down to a few options. If the infant has a normal ophthalmologic examination, including refraction, has no nystagmus and evidence of saccadic eye movements, DVM or cortical visual impairment (CVI) are the main choices. CVI is often associated with some history of perinatal event such as perinatal hypoxia, cerebral vascular accident, meningitis and acquired hypoxia. In one study, seizures were associated with 53% of cases and cerebral palsy with 26% [32].

However, if no history of obvious perinatal injury or no other neurologic symptoms are present, MRI may be needed to differentiate between CVI and DVM.

Etiology of isolated DVM

Initially, the main hypothesis regarding the etiology of DVM involved a possible delay in myelination of the optic nerve. This dates from the early descriptions of DVM infants with gray optic nerves [2–4]. During a fetus' and infant's maturation, myelination occurs from the brain and proceeds distally towards the eye. A postmortem examination of 18 infants by light and electron microscopy showed that myelin in the optic nerve near the globe started to appear at term and became fully developed by the age of 7 months [33].

Although the continued myelination of the optic nerve during early infancy may theoretically cause some visual symptoms, it is unclear whether this occurs in practice. Although little consensus has been established regarding VEPs and infants with DVM, several studies show normal responses when adjusted for age [12,16]. Thus, functionally, delay in myelination probably represents an insignificant contribution to the visual innatention symptoms of DVM. In addition, MRI studies of the optic chiasm and nerve suggest no difference between DVM patients and controls [18].

When considering the possible etiology of DVM, we should consider all the pathways that need to be working in order to achieve normal vision with fixation and follow. The sensory, motor and attentional mechanisms all need be functioning in order to provide a normal response to a visual stimulus by the infant.

The afferent visual system involves the sensory visual experience from the retina, optic nerve and visual pathways throughout the brain. There is little evidence of malfunction in this system. Patients with DVM show normal results on ERG and fundoscopic examination is age appropriate, making macular immaturity an unlikely cause. Additionally, neuroimaging of the optic nerve are within control groups [18]. The general absence of nystagmus shows a lack of anterior visual pathway disease. More importantly, normal results for tests such as vernier acuity and VEP, suggest an afferent pathway that is intact from start to finish [16].

The efferent (motor) visual system also appears intact as the brainstem saccadic generator is functional when measured. Studies have demonstrated normal age-related OKN in infants

with DVM [21]. In addition, other studies have demonstrated normal VOR with fast phase, consistent with age-specific patterns for infants [11,12,20]. Thus, despite ability to fix and follow, these infants display normal motor function when measured by other proxies.

Many authors have attempted to explain a possible mechanism for DVM based on their results. Lambert *et al.*'s review of infants with DVM led to the elimination of myelination of the visual system and macular immaturity on the basis of VEPs and ERGs [12]. In addition, they believed that the striate cortex was not involved, based on the normal pattern VEPs in the literature. Based on this, they suggested visual inattention originating from immaturity of the visual association area [12].

Tresidder *et al.* speculated that the cause for DVM may be a defect in subcortical extrageniculostriate visual system in infants [9]. This system may be damaged in some infants owing to hypoxia or some other insult, but visual symptoms may improve as the cortical system develops. The timing of visual recovery in infants with DVM may correspond with the development of other functions (binocular vision and smooth eye movements) that are dependent on cortical development [34].

Cocker *et al.* reported a case of identical twins, one of whom had type Ib DVM and was found to have abnormal acuity card procedure and grating pupillometry [35]. The authors concluded "that although the underlying defect is primarily subcortical, secondarily it delays the emergence of cortically mediated responses [35]."

Other authors believe that evidence of temporo-nasal OKN suggest that the geniculostriate system is intact in infants with DVM [21]. These authors suggest a delay in the development of extrastriate cortical structures involving an abnormality in attentional pathways or figure-ground segregation [21].

There is an abundance of information in the visual world and certain processes are in place to direct and focus our attention to any one part of the world. It has been shown that as a result of our brain's limited processing capacity, multiple stimuli compete for representation [36]. There are systems in place that bias towards the selection of one stimulus versus another. There is a bottom-up process that is a function of the stimulus' salience and there is a top-down mechanism that is influenced by attention centers in the brain [37].

The top-down attention centers derive from multiple areas outside of the visual cortex. The anterior system appears to help select the stimulus. Anatomically it includes the frontal eye fields and supplemental eye fields of the frontal cortex as well as globus pallidus, caudate, putamen and perhaps the posterior thalamus [5]. The posterior system also aids in attention and consists of the inferior parietal cortex, superior colliculus and pulvinar [5].

Although the exact pathways behind visual attention are not completely understood, it is possible that the mechanism behind DVM may be related to delayed development of this network in the brain. We need to also consider that DVM has multiple etiologies. However, these authors believe that dysfunction must exist in a common pathway to explain the symptoms.

Expert commentary & conclusion

Delayed visual maturation does not represent a single disease, but probably a symptom common to several neurologic abnormalities. Given the high correlation with prematurity and perinatal events, it is likely that isolated DVM represents a neurologic insult that is subtle and eventually overcome as the infant continues to develop. These insults may manifest in subsequent years as developmental problems, as seen in Hoyt's retrospective study [5]. Thus, we are dealing with a spectrum of neurologic disease with isolated DVM as the most benign and with the best prognosis.

The approach to the care of an infant with DVM should be interdisciplinary and coordinated among multiple pediatric specialties. Isolated DVM, with resolution of symptoms over time, has little long-term ophthalmologic significance. However, even those infants with isolated DVM should be carefully monitored by pediatricians, looking for any future signs of developmental delay.

Based on much evidence, it appears as though afferent and efferent visual pathways are intact in patients with DVM. It is possible that the initial delay is due to undetectable damage to the visual attention pathway. Thus, as Beauvieux first described, it may be more accurate to describe this phenomenon as a visual inattention rather than delayed visual maturation [1].

Five-year view: need for further study

Hoyt's prospective study of infants with DVM demonstrated a high incidence of neurodevelopmental delays [5]. However, a prospective study would be valuable in order to help us determine the true incidence of these neurologic developments so that we may better estimate prognosis. In addition, as suggested by Hoyt, a prospective study could identify sequelae of DVM that has not yet been discovered [5].

Key issues

- Delayed visual maturation (DVM) or visual inattention is a source of much anxiety for families of infants with this entity.
- DVM is classified as type I or isolated if it is not associated with any other neurologic or ophthalmologic abnormalities.
- Other classifications exist depending on whether neurologic, ophthalmologic or other conditions are also present.
- Isolated DVM patients have normal clinical examination, normal imaging and normal or near normal central eye movements.
- Prognosis of visual function in infants with isolated DVM is excellent with resolution of symptoms expected by 6 months of age.
- Prognosis in patients with other types of DVM are variable.
- Isolated DVM merits little long-term ophthalmologic attention, although developmental or other neurologic abnormalities may manifest with time.
- The etiology of DVM is unclear, although a delay in or injury to the visual attention centers are implicated.
- Further investigated is needed on the long-term sequelae of this entity.

If we hypothesize that DVM is due to some neurologic insult that is too subtle to identify, we should look for other possible causes for this injury. It has been well established that unconjugated bilirubin is neurotoxic at high levels and can cause such extreme damage in infants such as bilirubin encephalopathy. No studies have correlated the level of bilirubin levels with absence or presence of DVM. There is evidence to suggest that even moderate levels of hyperbilirubinemia can be neurotoxic to the developing infant [38]. An auditory processing syndrome has been described in children with elevated bilirubin [39].

A couple of studies have shown a possible, perhaps transient relationship between bilirubin levels and VEPs [40,41]. Studying a possible link between DVM and elevated bilirubin levels could provide insight on a possible mechanism.

The mechanism and site of damage for DVM is currently unknown, but study involving more advanced imaging modalities such as functional MRI could provide new insight. We could gain further clarity on which parts of the brain are affected. Further investigations such as those described could yet provide incredible insight in our understanding of this complex visual phenomenon.

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Activity Evaluation
Where 1 is strongly disagree and 5 is strongly agree

	1	2	3	4	5
1. The activity supported the learning objectives.					
2. The material was organized clearly for learning to occur.					
3. The content learned from this activity will impact my practice.					
4. The activity was presented objectively and free of commercial bias.					

1. Your patient is a 3-month-old girl who was referred to you by the patient's pediatrician. Her parents noticed that she does not fix her gaze on them or attempt to follow objects with her eyes. The patient has a normal birth history and appears to be otherwise healthy. What should you consider regarding the clinical presentation of type I delayed visual maturation (DVM) at this time?

- ☐ **A** Strabismus is often present during the stage of visual impairment
- ☐ **B** Convergent strabismus is more common than divergent strabismus
- ☐ **C** Patients rarely present with symptoms prior to 1 year of age
- ☐ **D** DVM type I rarely resolves prior to 1 year of age

2. Which of the following signs is most helpful in diagnosing this child with DVM?

- ☐ **A** Normal visual evoked potential
- ☐ **B** Abnormalities in the optic nerve at the optic chiasm
- ☐ **C** The inability of the child to fix her gaze and follow movement
- ☐ **D** Defects in multiple central eye movement systems

3. The patient's parents are concerned about other problems that might be related to their daughter's DVM. What should you consider regarding type II DVM?

- ☐ **A** Visual symptoms are often slower to resolve in type II DVM compared with type I DVM
- ☐ **B** Seizure control in type II DVM control has no effect on visual symptoms
- ☐ **C** Infants with type II DVM have congenital nystagmus and albinism
- ☐ **D** Type II DVM is associated with macular coloboma

4. The parents want to know why their daughter is affected by DVM. Which of the following defects appears most likely to explain the pathology of DVM?

- ☐ **A** Delay in myelination of the optic nerve
- ☐ **B** Defects in visual attention centers within the cerebral cortex
- ☐ **C** Defects in the afferent visual system
- ☐ **D** Defects in the efferent (motor) visual system