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A BLOWN PUPIL AND INTRACRANIAL HEMORRHAGE IN A 4-WEEK-OLD: A CASE OF DELAYED ONSET VITAMIN K DEFICIENCY BLEEDING, A RARE "CAN'T MISS" DIAGNOSIS

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☐ **Abstract—Background: Infants are at risk for vitamin K deficiency bleeding (VKDB) because of limited stores of vitamin K (VK) at birth and a low concentration of VK in human breast milk. Therefore, the administration of intramuscular (IM) VK at birth has been recommended since 1961 in the United States. Infants who do not receive IM VK and who are exclusively breast-fed are at increased risk for VKDB. While VKDB is rare, a common presentation of late onset VKDB is intracranial hemorrhage. Case report: We report the case of a 4-week-old infant who presented to the emergency department with lethargy and a grossly dilated right pupil. The parents denied trauma. A computed tomography scan revealed a right-sided subdural hematoma with midline shift. The infant's international normalized ratio was >10.9 and his prothrombin time PT was >120 seconds. VK was administered and the child was transferred to a tertiary care center for emergent neurosurgery. Why Should an Emergency Physician be Aware of This?: The difficult part of making this critical diagnosis is considering it. Any bleeding in a newborn without trauma should prompt inquiry regarding neonatal VK administration and a serum prothrombin time level. Fortunately, once the diagnosis is made, therapy in the emergency department can be lifesaving and is familiar to emergency physicians. Treatment parallels usual care for the adult with excess anticoagulation caused by warfarin. Prompt intravenous VK is universally accepted. Studies to support fresh frozen plasma or prothrombin complex concentrate are lacking but make good clinical sense for life-threatening bleeding.**

☐ **Keywords—bleeding; hemorrhage; newborn; treatment; vitamin K deficiency**

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INTRODUCTION

Previously referred to as hemorrhagic disease of the newborn, vitamin K deficiency bleeding (VKDB) is a rare and potentially fatal bleeding disorder of early infancy. There is a direct relationship between the omission of intramuscular (IM) vitamin K (VK) at birth and risk for VKDB; the relative risk for those children developing late onset bleeding is significantly greater than those who received IM VK at birth (1,2). Parental decisions to forgo VK at birth have been an issue (3,4). A recent spike in VKDB cases in Tennessee led to a joint investigation between the Tennessee Department of Health and the Centers for Disease Control and Prevention (5).

There are 3 accepted forms of VKDB (Table 1). From an emergency medicine standpoint, delayed onset VKDB is the form with highest prevalence of life-threatening intracerebral hemorrhage. Fortunately, timely treatment in the emergency department (ED) is not complicated and is potentially lifesaving. We present a case of

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Table 1. Vitamin K Deficiency Bleeding

Types of VDKB	Timing	Proposed Causes	Presentations
Early	≤24 hours	Maternal use of VK-inhibitory drugs, such as antiepileptics, warfarin, isoniazid, phenobarbital, and phenytoin (1,4–6,8,11); prophylactic VK at birth may not be protective (1,4,8)	Cephalohematoma, umbilical, ICH, intra-abdominal, intrathoracic, or GI bleeding (7,11)
Classic	2–14 days* (1,3–9)	Natural decrease in VK levels in the interim between placental stores and feeding source (5); normal transient decrease in VK-dependent factors, lack of needed bacterial flora in gut (3); poor placental transfer of VK and short half-life of liver stores (3); "mainly idiopathic" (11)	GI, nasal, skin, circumcision bleeding (4,6,11); ICH is rare (6); in 1 case series of 32 infants in Egypt, 21% had ICH (12); cephalohematoma and intrathoracic bleeding (7); ICH, GI, and other (1)
Late or delayed onset	1 week–6 months* (1,4–8); peak onset 2–8 weeks, may happen up to a year* (10); 1 week–"rarely beyond 3 months"* (9)	Primarily in exclusively breast-fed infants that did not receive IM VK at birth (8,11); malabsorptive diseases like cystic fibrosis, cholestatic liver disease, biliary atresia, and alpha-1-antitrypsin disease (5,6)	Vomiting, lethargy, pallor, decreased feeding, and seizure (4,10,12); "warning bleeds" nares, mucosa, and umbilicus (4,6); severe ICH or GI bleed (5); location of bleeding is intracranial 50% of time (4,7,11)

GI = gastrointestinal; ICH = intracerebral hemorrhage; VK = vitamin K; VKDB = vitamin K deficiency bleeding.
* Timing consensus is lacking for classic and late or delayed onset VKDB.

intracerebral hemorrhage caused by delayed onset VKDB in a 4-week-old male whose parents deferred IM VK at birth.

CASE REPORT

A 4-week-old male presented to the ED with a decreased level of consciousness. Both parents reported that the child was doing relatively well until the day before. At 8 PM of the previous evening, the child took 3 oz of breast milk as usual but then had an episode of projectile vomiting. The rest of the evening was uneventful and the child tolerated a few small feeds the morning of the presentation. The parents denied any fevers, rash, or cough. They presented to the ED for ongoing concerns that the child "was just not acting himself." The parents were attentive, concerned, and adamant that there was no physical trauma involved.

The child was delivered uneventfully 3 weeks early by cesarean section because of placenta previa. His birth weight was 2.7 kg and his weight on ED presentation was 3.6 kg. The mother reported that she declined routine IM VK at birth, and her rationale was that they opted not to have the child circumcised. The parents also declined routine immunizations.

The child was pale and ill appearing at the time of the physical examination. While bundled in his mother's arms, the child did not move or cry. When stimulated during the examination, he had a vigorous cry and moved all extremities. His rectal temperature was 35.9°C (96.6°F), his blood pressure was 109/63 mm Hg, his pulse was 112 beats/min, and his respiratory rate was 30 breaths/min. His breathing was unlabored and regular. His pulses where easily palpable in all extremities and capillary refill was normal. His cardiac, pulmonary, gastrointestinal, and genitourinary examinations were unremarkable. He opened his eyes spontaneously to stimulation, at which time a dramatically dilated right pupil was evident.

Immediate intravenous access was obtained and laboratory studies were performed. A computed tomography (CT) scan of his head revealed a right-sided subdural hematoma with midline shift (Figure 1). Scattered small subarachnoid hemorrhages were also present (Figures 1 and 2). The child's hemoglobin level was 6.1 g/dL. Coagulation studies were collected but the local laboratory was unable to correctly interpret the results, and these results were sent to the closest tertiary care hospital. Interventions in the ED included a 40-mL bolus of normal saline, 30 mL of uncross-matched O negative blood, and 1 mg of vitamin K.

Weather precluded air transfer, and a pediatric ground crew was sent from the accepting tertiary care hospital. The child's trachea was intubated before transfer and he was taken directly to the operating room at the accepting hospital. Coagulation results became available during transport; the child's INR was >10.9 and his PT was >120 seconds. The child underwent a craniotomy, a stay in the intensive care unit, and was eventually discharged home.