

Late Diagnosis of Hypoxic-Ischemic Encephalopathy in a Child with Normal Apgar Scores

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Abstract

M.S. was delivered at 40 weeks by emergent caesarian section after late decelerations were noted on exam. Her Apgar scores were 8 and 9 at one and 5 min, respectively. Although she had difficulty sucking a bottle, she was released from the hospital without neurological consultation. She went on to develop persistent physical and mental disabilities, including generalized hypotonia, impaired language development, and severe ADHD. The patient's symptoms were never definitively linked to perinatal distress, and the possibility of hypoxic-ischemic encephalopathy was not investigated. Her parents, deeply distressed by their lack of answers, brought her to our clinic as an 18-year-old. We ordered DW-MRI and found evidence of remote hypoxia. Paired with the clinical history, these findings suggest the patient suffered HIE due to fetal distress. This case demonstrates that overreliance on the Apgar score can lead clinicians to miss abnormalities in the critical window of early infancy and fail to connect later pathologies with birth trauma.

Keywords: Hypoxic ischemic encephalopathy; Developmental delay; Neonate; Infant; Children, Disability; Magnetic resonance imaging; ADHD

Introduction

First developed by pediatric anesthesiologist Virginia Apgar in 1952, the Apgar score has become a standard for assessment of the newborn [1]. Apgar is designed to provide a rapid assessment of a newborn's physiological state and identify the need for resuscitation. The score is calculated on the basis of five components [1]:

- Appearance: Color of skin
- Pulse: Heart rate
- Grimace: Reflex irritability
- Activity: Muscle tone
- Respiration: Breathing rate

Each component is graded on a 0-2 scale and the score is totalled to a maximum of 10. A score of 7-10 is normal, 4-6 is moderately depressed and 0-3 is severely depressed. A one-minute Apgar score greater than 7 usually indicates that the baby will require only routine post-delivery care [1]. Although the Apgar score can provide critical information about the newborn's immediate needs, it is a crude indicator of overall health [1]. Moreover, there is evidence that the subjectivity of some Apgar components can lead to disagreement and error in assigning a score [2-4]. A 2015 committee letter from The American College of Obstetricians and Gynecologists firmly states that "the Apgar score alone cannot be considered to be evidence of or a consequence of asphyxia, does not predict individual neonatal mortality or neurologic outcome, and should not be used for that purpose" [1]. Nevertheless, some practitioners rely on the Apgar score to provide a general sense of the infant's health and treat a normal

Apgar score synonymously with a normal infant. Overreliance on the Apgar score may lead clinicians to miss signs of pathology in the critical window of early infancy.

Case Presentation

M.S. was an 18 year old female brought to our neurology clinic by her parents because they were concerned about her station in life. The parents reported a long history of physical and mental disability. On inspection, M.S. had mild facial dysmorphism, nystagmus, and a prominent scoliokyphosis. On neurological exam, she was oriented to person and place but not to time. Her attention span and short-term memory were poor, and she had difficulty with speech, grammar, and reading. She was pleasant, but her social interactions were of a preadolescent level. On motor exam, she had spasticity in the upper and lower extremities with extremely poor dexterity of the upper limbs. There was diffuse hyperreflexia of all extremities.

A review of the patient's records seemed to indicate a static encephalopathy since infancy. M.S. was delivered by emergent caesarian section at 40 weeks of gestation after the OB/GYN noted persistent late decelerations. Her mother was 28 years old at the time of delivery and had reported no complications during pregnancy. The birth weight was 6 pounds and 14 ounces. The Apgar scores were recorded as 8 and 9 at one and 5 min, respectively. Given the normal Apgar scores, HIE was not suspected. Subsequently, although M.S. had some difficulty feeding, she was released from the hospital within 72 h without extensive workup or neurological consultation.

At home, M.S. continued to have difficulty sucking and required a nipple with an enlarged hole. In the first months of life, she displayed unusual flopping movements, impaired gagging and swallowing and a protruding tongue. She was consistently small for her age and failed to

meet developmental milestones. At 11 months, she was still unable to hold her head up, grasp objects, or sit up without assistance. She did not follow objects with her eyes when they were passed midline and showed no reaction to what her parents were doing.

At this time, M.S. underwent neurological consultation at her birth hospital. On exam, she was found to have generalized hypotonia, impaired cognition, and language deficits. Workup for thyroid dysfunction and organic acid disorders was negative. A standard MRI brain of the brain was unremarkable except for fluid in the left mastoid air cells. A full genetic screening was performed and revealed no abnormalities. No diagnosis was established at this time.

Despite extensive physical and cognitive therapy, the patient's deficits persisted. At 18 months, she could crawl with difficulty. She began walking with difficulty at the age of 2. At age 4, there were no signs of speech, social development was profoundly delayed, and motor deficits persisted. She was also diagnosed with severe ADHD. Her difficulties with language and concentration prevented her from attending regular classes, and she was enrolled in special education. She continued to require extensive therapy and constant monitoring by her parents or aides. Her condition was eventually characterized as a static encephalopathy with a low probability for improvement.

Although M.S. continued to receive follow-up care from the hospital at which she was born, the cause of her encephalopathy was never established, and she came to our clinic without a clear diagnosis. Her parents were deeply concerned about her ability to live an independent and fulfilled life and desperate for answers about the cause of their daughter's disability.

Given the patient's history of a complicated delivery and motor and cognitive deficits dating to birth, we considered that her disability could be the result of hypoxic-ischemic encephalopathy (HIE). However, this condition is usually identified in infants and is almost always associated with a low Apgar score. We ordered DW-MRI studies of the brain to look for evidence of HIE.

DW-MRI revealed hemosiderosis bilaterally in the mid and inferior cerebellar hemispheres. It also revealed mild prominence of the great horizontal fissure bilaterally and mild loss of volume in the superior semilunar lobules bilaterally, right inferior semiulular lobule, and lateral aspect of the left semilunar lobule. These findings were consistent with disruption of energy supply to the brain during a remote hypoxia. Given the clinical history, M.S. most likely suffered HIES due to perinatal distress. Her parents were informed of this probable diagnosis as well as the fact that her condition would likely require lifelong medical care, motor and cognitive therapy, and continuous supervision.

Discussion

This case demonstrates that a normal Apgar score does not definitively rule out HIE and should not be relied upon to establish a

newborn as healthy. It is important for physicians to recognize the limitations of Apgar. Apgar screening is designed to assess whether a newborn is physiologically stable and suggest the need for immediate interventions. It is not designed to assess congenital deficits or predict long-term outcomes. Moreover, the test relies on a number of subjective measures, limiting its reliability. In one study, 223 health professionals caring for newborns were given case presentations and asked to assign an Apgar score. Even pediatricians, who were the highest-scoring group, assigned the correct score only 68% of the time, while nurses in a community hospital setting were only 24% accurate [2]. In another study, neonatal and obstetric staff were shown recordings of 30 live births with an Apgar screening and asked to assign a score. Their scores differed on average more than 2.4 points from those assigned by staff present at the deliveries [3].

The limitations of Apgar screening have significant clinical implications. A recent study comparing different versions of the Apgar test found that the conventional test had a relatively low sensitivity for birth asphyxia (81%) [4]. Given that up to 20% of birth asphyxia cases may be missed on Apgar screening, practitioners providing follow-up care must consider HIE even in infants whose Apgar scores were normal. In this case, it is unclear whether the normal Apgar scores accurately reflected the patient's condition or were improperly assigned.

Conclusion

This case demonstrates the pitfalls of relying upon Apgar scores to establish an infant as normal. M.S. had difficulty sucking after a delivery characterized by late decelerations, but HIE was not suspected, and she was released from the hospital without an extensive workup. Subsequently, her physicians failed to link motor and cognitive deficits to complications of birth. It was only an MRI at 18 years of age that furnished evidence of HIE, providing her parents a much sought-after explanation. Better recognition of the limitations and proper use of the Apgar score may help to prevent such instances of missed diagnosis.

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