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Case report

Brain death in neonates: a case report

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The last ten years, the next ten years in Neonatology

Guest Editors: Vassilios Fanos, Michele Mussap, Gavino Faa, Apostolos Papageorgiou

Abstract

Brain death (BD) is the permanent and complete loss of cerebral and brainstem function. It is relatively uncommon in newborns with its percentage among deaths being 1-6.3%. BD leads to debate for medical, ethical and philosophical issues. It is a challenging condition in neonatal intensive care units (NICUs) since difficulties for BD diagnosis in neonates and ever more so in preterm neonates do arise. Revised guidelines for BD diagnosis definition include history with known etiology, clinical examination, apnea testing and neurological evaluation often assisted by ancillary tests.

We present the case of a near term female baby that was born with brain death due to hypoxic ischemic encephalopathy. We conclude that BD in newborns is a challenge to NICUs and there is a need for establishing and implementing new guidelines and checklists on national basis.

Keywords

Brain death, newborn, case report.

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Case report

A female baby born by Caesarean section due to decreased fetal movements at a gestational age of 36 weeks (BW 2,280 g) to a 42-year old gravida 3 para 3 mother. The baby born cyanotic, apnoic with heart beat rate < 60/min and an Apgar score 0 at 1 and 5 minutes respectively was intubated in the delivery room.

The mother was treated with magnesium sulfate during her pregnancy and she was on calcium and ferrum supplementation. Her pregnancy course was complicated with hypotension and anemia and had contractions 3 weeks before delivery for which she was placed on ritodrine. The neonate, admitted in neonatal intensive care unit (NICU) intubated, was assisted with mechanical respiratory support.

Laboratory tests were normal (complete blood count, acid base levels, metabolic profile, biochemical assay). Treatment with ampicillin, gentamicin was empirically initiated and it was discontinued 5 days later since blood cultures from day of life (DOL) #1 remained negative. Early-onset sepsis and TORCH screen test yielded negative results. Lumbar puncture was performed on DOL #12 and cerebrospinal fluid culture was found negative. Further examinations excluding metabolic abnormalities such as urea cycle disorders, lysosomal diseases, carbohydrate deficient glycoprotein, Niemann-Pick disease were also negative. Transcranial ultrasound was normal while magnetic resonance imaging/angiography showed hydropic degeneration of the hemispheres. Cerebral flow monitoring and electroencephalogram were performed and exhibited electrocerebral silence (ECS) and Doppler showed reduced brain blood flow. Newborn's clinical condition (comatose, apneic and with extensive hypotonia) remained unchanged from the time of birth up to DOL #16 on which she died still remaining under mechanical support. Absence of brain stem reflexes and pathological apnea test, twice evaluated 24 hour apart by two different examiners (neonatologists), led to the definite diagnosis of brain death (BD) at 38 weeks postconceptual age. Postmortem skin and brain tissues were attained and sent to the pathology department for examination.

Discussion

Brain death is defined as the biological event resulting in the permanent and complete cessation of the critical cerebral and brainstem function. Asphyxia, severe intracranial hemorrhage and infection account for the vast majority of the causes of BD in children and infants. Ethical dilemmas arise in NICU and the neonatologist usually is asked to work in moral and medical framework within which guidelines should be followed [1]. Defining BD in preterm neonates is a challenging issue in NICU since there have been difficulties in assessing brainstem function and level of consciousness in premature infants of gestational age < 37 weeks [2]. Recent literature has elucidated the definition criteria of BD that include a history with known etiology, clinical examination, apnea testing and neurological evaluation often assisted by ancillary tests [3]. Revised BD guidelines in infants require a thorough history by excluding known causes of coma such as hypothermia < 35°C, hypotension/ shock, metabolic disturbances and drug intoxication. BD mechanically supported neonates are comatose, apneic and lack brain stem reflex. An apnea test can be performed to ensure no spontaneous respirations after the PaCO₂ rises > 60 mmHg. Apnea test is demonstrated when the newborn fails to achieve spontaneous respirations in a hypercarbic state. Apnea test sometimes cannot be completed and neurological examination is unclear due to confounding variables such as systemic medication.

Additional tests are used to assist the diagnosis of BD. In newborns BD diagnosis is based both on clinical criteria and neurodiagnostic tests (electrophysiologic/brain blood flow). Ancillary studies have been used to complement clinical evaluation when there are uncertainties or difficulties in the neurological examination or apnea testing due to the newborn's condition. Four-vessel cerebral angiography, an invasive and time consuming test, has been traditionally the gold standard for the confirmation of BD. Additional tests have been developed to strengthen BD diagnosis including nuclear medicine cerebral perfusion test, electroencephalogram, transcranial ultrasound, magnetic resonance imaging/angiography, and evoked potentials [4]. There is a lack of specific guidelines for BD diagnosis in premature infants in NICUs in Greece [2]. In our unit, we use the revised diagnostic criteria by Nakagawa et al. proposed at their update of the 1987 Task Force Recommendations [3]. There seems to be an urgent need for establishment and implementation of new guidelines for BD in newborns on a national level.

Declaration of interest

The Authors declare that there is no conflict of interest.

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