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CASE REPORT

Ethical and medical management of a pregnant woman with brain stem death resulting in delivery of a healthy child and organ donation

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ABSTRACT

Maternal brain death during pregnancy remains an exceedingly complex situation that requires not only a well-considered medical management plan, but also careful decision-making in a legally and ethically delicate situation. Management of brain dead pregnant patients needs to adhere to special strategies that support the mother in a way that she can deliver a viable and healthy child. Brain death in pregnant women is very rare, with only a few published cases. We present a case of a pregnant woman with previously diagnosed multiple brain cavernomas that led to intracranial hemorrhage and brain stem death during the 21st week of pregnancy. The condition that can unequivocally be proven, using tests that do not endanger viability of the fetus, is brain stem death, diagnosed through absence of cranial reflexes. The patient was successfully treated until delivery of a healthy female child at 29 weeks of gestation. The patient received continuous hormone substitution therapy, fetal monitoring and extrinsic regulation of maternal homeostasis over 64 days. After delivery, the final diagnosis of brain death was established through multi-slice computer tomography pan-angiography. This challenging case discusses ethical and medical circumstances arising from a diagnosis of maternal brain death, while showing that prolongation of somatic life support in a multidisciplinary setting can result in a successful pregnancy outcome.

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Introduction

Brain death during pregnancy occurs rarely, while the delivery of a healthy child as well remains an even rarer event, with about 30 cases described in the literature to date, and only 12 viable infants delivered.¹ Management of a brain-dead pregnant patient needs to adhere to strategies that support the mother's condition, so as to permit delivery of a viable and healthy child, while protecting the fetus from adverse effects during that pre-delivery period.

This case report describes specific ethical and medical circumstances in confirming the diagnosis of brain death

in a pregnant woman. It is the first case in Croatia of confirmed brain death in a woman at 20 weeks of gestation and having obtained consent to do so, describes her management over 64 days, until delivery of a healthy child and subsequent organ donation.

Case report

A 34-year-old woman was admitted to the neurological department due to progression of left-sided weakness of her extremities and facial asymmetry, which had started 15 days before. On the day of admission, she was 20 weeks of gestation in her second pregnancy. She had been diagnosed with multiple cerebral cavernomas in the left cerebral peduncle, the right hypothalamus and the left thalamus two years previously; no invasive treatment had been recommended. Her pregnancy had been followed up regularly and appeared to be normal.

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After admission, the patient's Glasgow coma scale (GCS) was 12–13 (eye 3, verbal 4, motor 5–6), but over several hours worsened to GCS 3 (eye 1, verbal 1, motor 1) with preserved spontaneous respiration. Urgent brain magnetic resonance imaging (MRI) showed extensive hemorrhage in the right thalamus, mesencephalon and brain stem, with intraventricular hemorrhage, decompensated hydrocephalus and transtentorial and transforaminal cerebral herniation. During the MRI, the patient had a respiratory arrest, was resuscitated and transferred to the intensive care unit (ICU).

At this stage, an urgent obstetric ultrasound showed a viable fetus, with normal biodynamics and biophysical profile. Clinical testing to confirm the diagnosis of maternal brain death was performed twice. All cranial nerve reflexes were absent, with no response to atropine. The patient had no spontaneous respiration, but the apnea test was not performed due to possible deleterious effects of maternal hypoxia on the fetus. Confirmation of brain death by multi-slice computerised tomography (MSCT) contrast pan-angiography could only be confirmed after delivery.

After consulting the patient's family and obtaining informed consent from her husband, a multidisciplinary team including anesthesiologists, obstetricians, neurologists and neurosurgeons was assembled to plan medical management.

The Croatian criteria for brain death are defined by the Ministry of Health Rulebook for Diagnosing Brain Death (published 2004, amended in 2012), adhering to the European Parliament and Council Directive 2010/53/EZ and European Commission Directive 2012/25/EU.² Brain death occurs after an irreversible loss of cerebral, cerebellar and brain stem function. The diagnosis is made through two consecutive clinical examinations, and confirmed by one or more diagnostic tests by an anesthesiologist and a neurologist or neurosurgeon. The etiology of brain death must be well known and documented by a brain CT or MRI scan. The patient must be in an apneic coma with absent clinical reflexes, with a negative apnea test. Other reversible causes that can mimic brain death (hypothermia, hypotension, hepatic encephalopathy, hyperosmolar coma, pre-terminal uremia or alcohol or pharmacologic intoxication) must be excluded. To confirm brain death, one of the following must be used as a confirmatory test: brain pan-angiography, transcranial Doppler scintigraphy, perfusion radionuclide scintigraphy or MSCT contrast pan-angiography. Most European countries require only brain stem death to be diagnosed for a patient to become an organ donor, rather than cortical brain death, but the latter must be diagnosed in pediatric patients and in unclear cases associated with possible intoxication. Croatian law explicitly requires the patient to be diagnosed with brain death when considering organ donation. In addition, after a diagnosis of brain death is established,

therapeutic procedures are only allowed in cases when the patient is an organ donor.

The condition we could unequivocally prove through available tests, without endangering the fetus, was brain stem death, diagnosed by the absence of cranial reflexes. An apnea test or any other based on it, and radionuclide or ionizing radiation, were contraindicated due to the ongoing viable pregnancy. At the request of the patient's husband, and with approval from the University Hospital Center's Ethical Committee, a conservative approach was adopted. Measures necessary to maintain maternal homeostasis were implemented due to fetal immaturity, and informed consent for organ donation was obtained from next-of-kin should brain death eventually be confirmed. The decision was made to continue somatic support until the patient reached 32 weeks of gestation and for cesarean delivery (CD) to be performed.

No special interventions apart from regular fetal cardiotocography (CTG), biophysical profile monitoring and betamethasone therapy were needed during the pregnancy. All other therapeutic interventions were tailored to avoid administering substances harmful to the fetus, but appropriate in the treatment of recurrent pneumonia and sepsis. Monitoring consisted of repeated electrocardiograms (ECGs), invasive arterial and central venous pressure monitoring and urinary catheter content measurements. She developed hemodynamic instability with marked hypotension, diabetes insipidus and diabetes mellitus. A continuous infusion of norepinephrine for vasoactive support was used, with desmopressin added intermittently to control diabetes insipidus. A continuous infusion of short-acting insulin was used for diabetes mellitus treatment, along with methylprednisolone. Levothyroxine was given intravenously. Obstetrical examination and ultrasound were performed every two days, and fetal heart monitoring was performed daily from 26 weeks of gestation onwards. Betamethasone therapy was administered to improve fetal lung maturity and reduce the risk of fetal respiratory distress syndrome.

The patient developed pneumonia twice (caused by *Haemophilus influenzae* the first time and by *Pseudomonas aeruginosa* the second) and developed signs of sepsis on three other occasions without underlying pneumonia (caused in turn by *Enterococcus* species, *Enterococcus* species and *Acinetobacter Baumannii*, and by *Pseudomonas aeruginosa*). The patient was treated with antibiotics according to the microbiological findings of blood cultures and tracheal aspirates. Particular care was taken to avoid antibiotics that might harm the fetus. Prophylaxis against gastric ulceration and thrombosis was administered, hypothermia was countered with heating blankets and physical therapy was performed daily. The patient was fed enterally. Due to the need for prolonged mechanical ventilation, a percutaneous tracheostomy was performed.

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