CASE REPORT

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Ethical and Legal Dilemmas Around Termination of Pregnancy for Severe Fetal Hydrocephalus, Spina Bifida Aperta and Meningomyelocoella

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ABSTRACT

Introduction: There are many ethical and moral dilemmas regarding the termination of pregnancy(TOP) with severe fetal anomalies. **Aim:** Our aim is to present a case of severe fetal hydrocephalus (HCP), spina bifida aperta and, meningomyelocoella (MMC). **Case report:** A gynecologist examined a 23-year-old patient with vital pregnancy of 24/25 week of gestation (WG) with the anomaly of the fetus. At the Perinatological Medical Advisory Board, a decision was made that the pregnancy should be continued and monitored bearing in mind that pregnancy exceeded the legal framework for TOP. Medical Advisory Board's ultrasound examination showed the following: severe hydrocephalus (HCP), spina bifida aperta, hyperehogen intestine, pes equinovarus. Via multidisciplinary consultation it was decided to make a delivery with the elective caesarian section (CS) causing as little trauma to the fetus as possible, with 37 WG completed due to the pelvic presentation and fetal anomalies. The patient gave birth via CS to a live female newborn—birth weight 3920 grams, birth length 56 cm, head circumference 48 cm, and Apgar score 8/8. The head was hydrocephalic with spaced suture. There was thoracolumbar defect of spina bifida aperta and meningomyelocele (MMC) 10x12 cm in size. An urgent surgical procedure - the external ventricular derivation of the liquor, and then the successful resection and plastic meningomyelocele was performed by a team of neuro and plastic surgeons. During the fourth postoperative day due to a suspicion of abscess collection and febrility of the mother a relaparotomy is performed and the abscess collection of Retzius space was found. Due to the fall in blood count, blood transfusion in a total dose of 580 ml was given. The patient was discharged on a home treatment as she had a regular general and local status. After the surgery, the condition of the newborn resulted in deterioration, the progression of HCP and dehiscence of head wounds and the thoracolumbar region, in spite of all the measures taken. At that time a decision was made to provide palliative care and this decision was conveyed to the mother. The child had a prolonged apnoeic episode and was not resuscitated. The child died in hospital after surviving for two months postpartum. **Conclusion:** Indication of TOP based on fetal anomalies could be a medical decision but also a personal moral choice of the mother related to legal rules, socio-cultural values and religious beliefs.

Key words: Fetus as a patient, Fetal anomalies, Delivery, Ethics, Termination of pregnancy.

1. INTRODUCTION

The prevalence of meningomyelocoella (MMC) has a world wide range of 1.7- 63.9 per 10 000 live births. The reported hydrocephalus (HCP) prevalence ranges from 2.5-8.2 cases per 10 000 live births (1, 2, 3).

HCP occurs in approximately 0.05 to 0.14 % of fetuses and accounts for about 12 % of all serious anomalies at birth. In 70 to 86% HCP is accompanied by other major anomalies: these include heart, brain and, in about one third of cases, MMC (3).

Prenatal detection rate of congenital HCP was 61% in study Garne et al. (2, 4).

Johson et al reported that TOP was more common when prenatal diagnosis was made less than 24 WG rather than later (86 vs. 27%) (5).

TOP is reported from 48% to 50% of HCP and in ~63% of spina bifida cases based on individual studies ranging from 31 to 97% (1, 5).

Prognosis for fetuses with congenital HCP was poor. One European study in 2010 showed that 47% of all cases were live births and of these 25% died within the first week after birth and at 1 year of age mortality was at least 38% (2, 6).

The advances in diagnostic imaging technology have increased

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the ability of early prenatal diagnosis of fetal HCP and MMC which is crucial for management (1, 7, 8). 3D and 4D examination moves prenatal diagnosis to the earliest possible gestation, which means that 11–14-week scan becomes the first mini-anomaly scan to diagnose and assess severe structural abnormalities (9). Doctors have legal, moral and ethical obligations to provide an accurate antenatal diagnosis and to inform parents about neonatal prognosis with anomalies (7).

If the diagnosis is suspected we should wait for the results of invasive testing to bring the final information to parents (6).

Prenatal diagnosis of HCP and MMC allows the parents to be counseled for prenatal/postnatal intervention, or TOP. This can create difficult discussions and decisions for patients and clinicians in predicting the prognosis and the course of the affected pregnancy (1).

Many countries do not distinguish between lethal and nonlethal abnormalities and there is no normative list of what can be considered "severe fetal abnormality" that would satisfy the criteria for TOP, leaving difficult decisions regarding TOP to parents and clinicians (4, 7, 9, 10).

A recent study has revealed no reported cases of pregnancy and childbirth of severe fetal hydrocephalus with spina bifida aperta occurring together with meningomyelocoella.

2. AIM

We present a case of severe fetal HCP, spina bifida aperta and MMC.

3. CASE REPORT

Antenatal history

A gynecologist examined a 23-year-old patient for the first time because she noticed that her belly was growing and she felt the fetal movements. She said that she had regular menstrual cycles for the previous four months, only the menses were shorter and less abundant. There were no peculiarities in personal and family history, except for tobacco abuse. The gynecological clinical and US examination confirmed 24 weeks and 4 days of gestation (WG), and a vital pregnancy with anomaly of the fetus. At the Perinatological Medical Advisory Board a decision was made that the pregnancy should be continued and monitored bearing in mind that pregnancy exceeded the legal framework for TOP. The Perinatological Medical Advisory Board's US examination: Fetal dynamics: neat, fetal heart activity: +, breech presentation, Fetal biometrics: biparietal diameter: 6.83, head circumference: 24.16, ventricular-head ratio: 0.88, abdominal circumference: 20.86, femur length: 4.82, estimated fetal weight: 863 grams. Severe hydrocephalus, spina bifida aperta (1,7 cm), hyperechogenic intestine, pesequinovarus (Figure 1, the first row) were diagnosed. All laboratory findings of the patient made during the pregnancy were in reference values, including microbiological analysis of vaginal and cervical swabs. By multidisciplinary consultation consisting of 3 gynecologists, subspecialists of fetal medicine and obstetrics, pediatric surgeon, neurosurgeon, paediatrician subspecialist neonatologist, and a lawyer, because of the pelvic presentation and fetal anomalies, the delivery was decided to end with the elective caesarian section - causing as little trauma to the fetus as possible, with 37 WG completed. The patient came urgently because of the preterm rupture of membranes and leakage of amniotic fluid, she did not bleed, she felt no pain, and the movements of the fetus were regularly felt. In the gynecological examination, evidently amniotic fluid was leaking, the cervix was shortened, the dilatation was 4 cm, breech presentation, fetal heart beat was regular, and the contractions were irregular. Cardiotocographic records were physiological.

Neonatal history

Laparotomy secundum Joel-Cochen and caesarian section secundum Misgav-Ladach were performed. Due to the inability of the extraction of the head, the T cut on the uterus was made and a live female newborn was born weighted 3920 grams, birth length 56 cm, head circumference 48 cm, and Apgar score 8/8. The head was hydrocephalic with spaced suture. There was thoracolumbar defect of spina bifida aperta and MMC size 10x12 cm with the diminished structure of the neural ridge (because of the pronounced displaceability of neural masses anatomical differentiation was not possible) (Figure 1, the second row). She had spontaneous movements of the upper extremities, the lower was paretic, and both feet in the calcaneovalgus.

Postnatal history

Echocardiosonographic findings indicated an aneurysm-altered interatrial septum. Lungs X-ray indicated bilateral basal shading and possible inflammatory infiltration. An emergency CT brain pointed to high-grade hydrocephalus and an enlarged chamber system without a clear differentiation of its boundaries (Figure 1, the second row). An urgent surgical procedure was performed by the team of neuro and plastic surgeons which included the external ventricular derivation of the liquor, and then the successful resection and plastic MMC in the thoracolumbar region. The newborn had stable vital parameters, pulse rate: 146 / min., O2 saturation 97%, and respiration rate: 56 / min.

The fourth postoperative day due to suspicion of abscess collection and febrility of the woman, a relaparotomy was performed and the abscess collection of Retzius space was found. After the toilet and drainage, antibiotic therapy was administered recommended by a clinical pharmacologist. Due to a fall in blood count, blood transfusion was given in a total dose of 580 ml. She was discharged for home treatment.

After the surgery, the condition of the newborn resulted in deterioration, the progression of hydrocephalus and dehiscence of wounds on the head and the thoracolumbar region, in spite of all the measures taken.

At that moment, a decision was made to provide palliative care and this decision was conveyed to the parents. The child had a prolonged apnoeic episode and was not resuscitated. Baby died in hospital after surviving for two months postpartum.

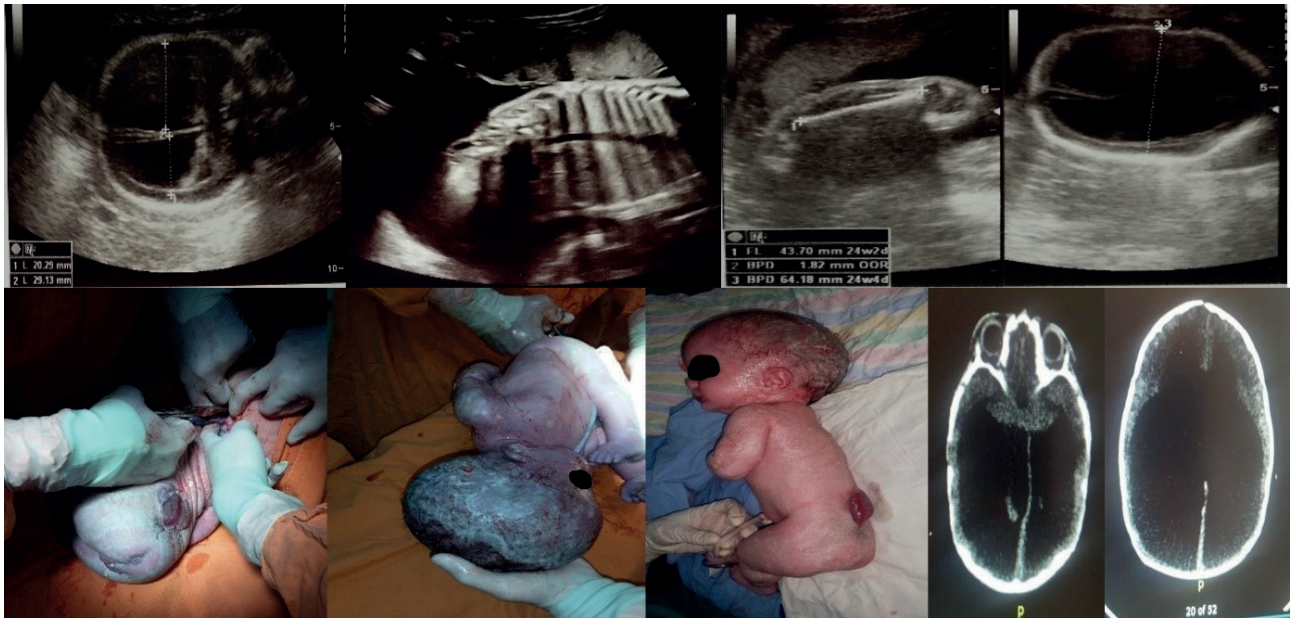

Figure 1. First row: Ultrasound examination at 24/25 WG. Second Row: The newborn with severe fetal hydrocephalus, spina bifida aperta and meningomyelocoella, and severe neonatal hydrocephalus on CT of the head

4. DISCUSSION

The case presented illustrates some of the ethical dilemmas around management of such clinical cases - here we have a woman who had just found out that she was pregnant and was faced with the fact that she was carrying a fetus with anomalies. In addition to two shocking news, she faced the fact that the pregnancy exceeded the time frame for termination of pregnancy (TOP). She was overwhelmed by psychological and physical fatigue, and the conflicting emotional and moral dilemmas associated with raising a physically challenged child while taking care of her family. She was eventually relieved by the natural outcome of early neonatal death.

A woman who was initially shocked by the news, learned to bond with her severely physically challenged baby as the pregnancy progressed, though the outlook was that of a shortened, poor quality life with severe neurocognitive deficits.

In countries with liberal abortion laws, TOP is allowed to women of any age within the first 12 weeks of gestation without restrictions. In countries with tighter abortion laws, TOP due to fetal anomalies can expose doctors and the patient to possible conflict and prosecution. In late trimester TOP is allowed where the pregnancy is likely to endanger the woman's life, the fetus is severely malformed, or there is a risk of severe injury to the fetus (7).

There are two extreme diversities, some countries have strict laws that forbid TOP, whereas other provide almost total freedom for both families and clinicians when the procedure is preferred (11, 12).

In the worldwide practices, although the gestational age limit (24 weeks) seems to be a legal restriction, it also has a scientific basis (11, 12). The fetal viability threshold, which is still described as 22-24 weeks of gestation, but nature of anomalies makes fetuses 'non-viable' at any gestational age (11, 12, 13).

However, as shown in our case, such decisions must be based on universally applicable criteria taking into consideration local constraints.

Delays in diagnosis and missed opportunity have contributed to considerable suffering for both parents and the affected children, and has the potential to culminate in maternal morbidity or even mortality (8).

Johson et al reported that greater frequency of TOP is expected at earlier gestational ages because many regions have laws restricting the gestational ages at which TOP can be performed (5).

The only published scientific guide related to TOP is a study by the Royal College of Obstetricians and Gynecologists, which was revised in May 2010 (12, 13).

All stuff involved in the care of a woman or a couple facing a possible TOP or continuation of the pregnancy with palliative care of the infant must adopt a non-directive, non-judgmental and supportive approach (1, 7, 10, 11, 12, 13).

The legal rules, beliefs, emotional needs and sociocultural values of the family should be in the center of the counseling process taking into the account the best interest of the child regardless of disability and neonatal outcome (7, 11, 12, 13, 14).

Doctors are under a legal obligation to counseling parents about the risk of a fetal anomaly, diagnosis, prognosis, and the treatment so that they can make an informed decision regarding TOP. The physician should discuss the options and potential complications which may occur to the fetus and pregnant mother (3, 4, 7, 8).

Decisions to perform TOP need to be undertaken by an institutional committee formed by the institution, and may be overseen by a national committee to ensure that the issues pertaining to TOP are performed in a way which would effectively avoid mistakes (8).

The physician should support women choice and offer medically reasonable alternatives, but not make any recommendations pro et contra alternative (3, 9, 10).

Therefore, a woman should be given the choice between TOP or continuing her pregnancy, resulting in the birth of a physically challenged child with severe physical or neurodevelopmental disorders, regardless of the physician's personal views (4, 5, 7, 9, 10, 12).

In our case, except legal restrictions, the mother continued her pregnancy because she believed in her baby's right to live.

A pregnant woman is in double role from the ethical point of view – a parent who must make a decision for the health of her future child, but also as a patient who decides for her own health (4, 5, 7).

Other moral dilemmas involved in such cases are further illustrated by our case, where the parents initially demanded TOP, but when this failed, they grew to love and care for the child despite its disability.

In these situations women's rights to autonomy and rights of the fetus as a patient can create conflicting moral dilemma regarding TOP and clinical management of neonates with severe congenital anomalies, as well as doctors' duties towards society (7, 11).

If the parental wishes are contrary to the infant's well-being, they can be legally overridden in the fetus's best interest because the doctor has the obligation to protect those interests (4, 7).

By favoring maternal and fetal interests physician's legal risks should not be overlooked, because doctors must be guided by ethical principles but also by local and international laws (3, 7).

Fundamental ethical principles in the care of malformed fetus pregnancies are the benefits for the patient and respect of the patient autonomy (4).

Islamic countries allows TOP if the pregnancy has not progressed beyond the 120th day of pregnancy, which is also referred to as the day of ensoulment (8).

Polyhydramnios can cause significant problems: maternal discomfort, placental abruption, unstable fetal position, ineffective uterine activity during the labor and postpartum haemorrhage (6). Our patient had polyhydramnios but did not experience any of the above complications.

The idea of promoting a fetal surgery that would potentially benefit the fetus in selected cases of fetal malformation, sometimes even increasing maternal risk, involves a pool of cultural, religious, legal, and technological factors and many conflicts and debates between specialized international organizations (4, 15, 16).

Placement of a ventriculo amniotic shunt in utero is possible; this approach has had limited success, and is currently regarded as experimental (3).

In some countries and in certain conditions cephalocentesis promotes the well-being of the hydrocephalic fetus (3, 10).

There is a question, what is the safest method of birth in patients with fetal anomalies like HCP and MMC? The alternative approach involves continuing the pregnancy until the labor begins spontaneously, followed by vaginal delivery (3).

Women with informed consent, can authorize and refuse caesarian delivery, and face the physician with a significant and challenging ethical conflict (3, 9, 10).

Women must know that there is a possibility, especially with caesarian section, of infant survival with permanent handicaps, depending on the severity of the hydrocephalus and the presence of other anomalies (3).

This approach attempts to minimize fetal injury and the exposure of women to the risks and complications of surgical delivery, with an increased rate of mortality and morbidity (3, 6, 14).

Caesarian premature delivery is indicated in the fetal malformations that require immediate surgical correction in a sterile environment (severe hydrocephalus, ruptured myelomeningocelus), but there is no unique recommendation (1, 4, 17, 18).

In our case, the patient had a severe infection as a complication of the caesarian section, so she consequently had a relaparotomy and extended hospitalization with all the accompanying consequences.

Cardiotocography is not routinely advocated during the labor and women should be appropriately counseled in this regard (6).

A multidisciplinary team approach is the ideal individualized management of patients with fetal anomalies, involving obstetricians, neonatologists, sonographers, bereavement and loss midwives, social support services and use of perinatal hospice care (6, 7).

If a postmortem examination is declined then a placental or cord biopsy should be sent for cytogenetic analysis and DNA storage, X-rays, computerized tomography or magnetic resonance imaging, and geneticist review according to local protocols (14).

Subsequent pregnancies may need to be managed by a fetal medicine specialist with anomaly ultrasound and/or invasive prenatal diagnostic techniques. It may also be necessary to refer the family to a clinical geneticist for genetic counseling and/or testing (14).

The birth of a severe disabled neonatus is an additional burden for the already limited health resources in poor countries (7). Moral obligations on clinicians is to prevent the waste of scarce healthcare resources (7). TOP for fetal abnormalities are often done to minimize burdens on parents, society, communities, institutions, and health care professionals (7, 8, 9, 10).

In resource poor settings like in most southeastern Europe countries, decisions regarding the use of scarce healthcare resources are sometimes paternalistic (17, 18).

5. CONCLUSION

The case we reviewed here was one of many encountered during the clinical practice at Southeastern Europe hospitals. The case illustrates some of the conflicting ethical and moral dilemmas surrounding management of liveborn neonate delivered following exceeded term for TOP for antenatal diagnosed hydrocephalus and spina bifida with meningomyelocoella, respectively (17, 18).

This case report and literature review aims to provide a framework for the management of pregnancies complicated by fetal anomaly.

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