Life and Medical Ethics in Pediatric Neurosurgery

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Abstract

Ethical issues in the field of pediatric neurosurgery, including prenatal diagnosis, palliative care for children with an intractable serious disease, and medical neglect, are discussed. An important role of medicine is to offer every possible treatment to a patient. However, it also is the responsibility of medicine to be conscious of its limitations, and to help parents love and respect a child who suffers from an incurable disease. When dealing with cases of medical neglect and palliative care for an incurable disease, it is critical to diagnose the child's condition accurately and evaluate the outcome. However, to treat or not to treat also depends on the medical resources and social-economic status of the community, the parents' religion and philosophy, the policies of the institutions involved, and the limits of medical science. Moral dilemmas will continue to be addressed as medical progress yields treatments for untreatable diseases in the future.

Key words: medical ethics, pediatric neurosurgery, prenatal diagnosis, selective treatment, medical neglect

Introduction

In the field of pediatric neurosurgery, I have sometimes encountered ethical problems. At the 74th annual meeting of the Japan Neurosurgeons Society, ethical issues were discussed comprehensively for the first time. Ethical problems in pediatric neurosurgery can be associated with prenatal diagnosis, palliative care for children with intractable serious disease, and medical neglect. In this paper, I will present some ethical problems I have faced in my clinical career, and how they were addressed.

Ethics in Prenatal Diagnosis

The prenatal diagnosis of central nervous system (CNS) malformations has been increasing due to improvements in diagnostic technology. However, early in fetal development, the morphological diagnosis of some CNS malformations is difficult, and the long-term outcome is still unclear. Therefore, it can still be challenging to make an early diagnosis for prenatal counseling.

In 2012, we formed a multi-disciplinary prenatal support team (MDPST) at our hospital. This team includes obstetricians, pediatricians, neonatologists, pediatric neurosurgeons, nurses and midwives from an outpatient clinic, neonatal intensive care unit (NICU), growing care unit (GCU), and maternal

fetal intensive care unit (MFICU) staffs, physical therapists, clinical psychotherapists, medical social workers, and medical department secretarial staffs. The team meets every 2 weeks. We also introduced a full-time perinatal coordinator position, the first in Japan. This coordinator guides and informs the parents, coordinates the related staff, and manages the MDPST meeting. To date, our MDPST has supported over 126 families.

After an accurate diagnosis is made, the MDPST provides three kinds of support. The first is for cases in which the symptoms can be improved by treatment and the child can return home and spend quality time with the family, even if a disability remains. In these cases, we will discuss the time and method of delivery and how to treat the child after birth, and provide comprehensive support for the parents, including explanations to facilitate their acceptance of the child. In some cases, the parents will make a prenatal visit to the NICU, where the baby will be placed after birth. Second, in cases of multiple malformations, untreatable heart or respiratory disease, or untreatable severe brain anomalies, we provide selective or palliative treatment while withholding or withdrawing life-prolonging medical treatment. The third type of support is for the termination of pregnancy (TOP) decision. In Japan, diagnosis before 21 gestational weeks may result in TOP because of maternal conditions. Even in such cases, the decision can be difficult for parents. Often

when a fetus has a serious disease, the pregnancy is continued after the parents learn about and accept the fetus' condition. Regardless of the situation, when TOP is a possibility, the team provides honest and sensitive support.

Case 1

A 38-year-old female was pregnant for the first time. She was referred to our hospital because the male fetus was found to have multiple malformations, including acrania, congenital cystic adenomatoid malformation (CCAM), and radial hemimelia at 17w3d. The parents decided to terminate the pregnancy after repeated discussions, and the prenatal coordinator made the arrangements. After delivery, the nurses of the MFICU decorated the child's dead body with flowers and pretty clothes that had been made by hospital volunteers who had also lost a baby. They gave him a letter written by a volunteer group called the Mermaid Boutique that said, "We who have lost children just like you, sewed these clothes stitch by stitch from our heart for you. We also sewed them for ourselves, to help us recover from our deep sorrow. Please play with other children wearing these clothes when you meet them in Heaven. We speak to our angels while sewing. Fathers, mothers, and children, you are not alone." After TOP, the mother is sometimes depressed due to feelings of loss and guilt. The mother in this case also joined the Mermaid Boutique, and delivered her second baby in our hospital.

Palliative care for children

How to treat children with a serious disease has long been a topic in the field of pediatrics.¹⁾ A guideline working group for children's terminal care in the ethical committee of the Japan Pediatric Society produced a document called, "Guideline for the discussion of medical care for children with serious disease" in April 2012 (http://www.jpeds.or.jp/uploads/files/saisin_120808.pdf). Progress in medical technology has improved our ability to prolong life. The aim of this guideline is to make sure the conversation between medical staffs and family members of a newborn with serious disease affirms the rights of the baby.

However, for a child with a serious disease who can expect only a poor prognosis, an endless fight for life can result in physical suffering for the child and distress for the family. While a mature adult has acquired a view of life and death through experience, a child has not, so decisions about the child's life are greatly dependent on the parents' wishes. However, the parents' wishes are not necessarily the same as the child's or in the child's best interest. The ethical committee of the Canadian Pediatric

Society states, "Children's best interest can only be achieved by comparing the benefit achieved by treatment administered under certain policy and the danger and suffering inflicted by that treatment."

When the disease is serious and only a poor prognosis can be expected, we will disclose that fact to the family, with the understanding that treatment is completely optional in such a case. Advance care planning to determine what can be done for the child after birth is conducted. It is important to assess what is best for a child with a poor prognosis, and to consider terminal care when appropriate, to protect the human rights and dignity of children who cannot speak for themselves.

Case 2

A fetus with 13 trisomy had semilobar type holoprosencephaly, severe heart disease, and a cleft lip and palate (Fig. 1). We had many conversations within the MDPST and with the parents before the child's birth and agreed not to conduct any prolongation of life. The baby did not enter the NICU, and spent time in the mother's bed with visits from the family. The baby passed away 4 days after birth, in the presence of the mother and family.

Case 3

A fetus with a chromosomal abnormality had lissencephaly and hypoplastic left heart syndrome. We had many conversations before the child's birth, especially between the parents and pediatric

Fig. 1 Fetal MRI of case 2 shows semilobar type holoprosencephaly.

cardiologists, and agreed not to conduct any prolongation of life. This baby also did not enter the NICU and stayed with the mother. The baby was discharged as the symptoms stabilized. The doctor in charge of the NICU made several urgent visits to the baby's home, but the baby was able to spend about 30 days with the family and passed away 49 days after birth.

Medical neglect

Medical neglect is a parent's refusal of medical care for a child, even for a curable disease. Clinicians need to be aware of medical neglect and seek to avoid it.

Case 4

In a male fetus, ventriculomegaly and occipital encephalocele were diagnosed at 34 gestational weeks. Our team planned to repair the encephalocele and to perform ventricle-peritoneal (VP) shunt after selective caesarean section. However, when the parents were informed, they refused all of the surgical interventions due to their religion. We were able to plan for the baby to be delivered by spontaneous vaginal delivery, because the encephalocele and head circumference were not excessively large. The baby was born by spontaneous vaginal delivery, with a body weight of 3740 g and a skin-covered encephalocele in the occipital region. computed tomography (CT) and magnetic resonance (MR)

images showed ventriculomegaly and that the encephalocele was occupied by cerebral spinal fluid (CSF) and brain tissue (Fig. 2). The baby had no other CNS or general malformations.

A week after birth, the baby developed vomiting and apnea, and showed progression of the ventriculomegaly and an increasing head circumference. At this stage, optimal treatment would deliver a good outcome. Our team informed the parents that without treatment the child would develop increasing physical and mental retardation. The parents still refused the surgery due to their religious beliefs. They believed that surgery would violate the child's body, which was borrowed from God, and cause his soul to be rejected by God. They stated that their sin appeared in their child's body as a disease, and that they would cure his disease with prayer. We presented this situation to the Medical Review Board of the Osaka National Hospital as a case of child abuse and medical neglect, because the optimal treatment would bring a good outcome, and no treatment would cause the child to develop retardation. We then placed this case in charge of a child guidance center.

A representative from the child guidance center appeared before a family court to request suspension of the parents' parental rights and select a substitute person to give consent for the baby's surgery. The family court ordered the suspension of parental rights and the selection of the substitute, professor

Fig. 2 Preoperative images of case 4. Left; CT scan shows ventricular dilatation. Right; CT scan shows that the encephalocele, 3 × 4 cm in size, is filled with CSF and brain tissue.

emeritus Satoshi Matsumoto. The court stated that surgical treatment for the child was necessary and appropriate from a medical standpoint, and that the parents' refusal was harmful to the child, even though it was based on their religious faith. That is, the parents' refusal of surgery was considered child abuse. The child needed to undergo the surgery as soon as possible.

Thereafter, the encephalocele repair and VP shunt were performed under the consent of Dr. Matsumoto. The suspension of custody was then withdrawn, and the child was discharged to his home.²⁾

Discussion

Ethics in prenatal diagnosis

The management after prenatal diagnosis of a CNS malformation varies widely depending on the fetal conditions. For some diseases, intrauterine treatment is very effective, with good outcomes. The manifesto of the Fukuoka Society of the Fetus as a Patient, adopted in 2004, states, "Doctors, other medical workers, and society as a whole have a sincere obligation to provide adequate diagnosis and treatment to the patient fetus. Scientific assessment procedures, social recognition, and new fetal treatment and management methods should be applied the same as for children and adults. In the diagnosis and treatment of the fetus, the mother's human rights and decisions should be fully respected." On the other hand, cases in which optimal treatment is still predicted to result in a severely disabled outcome may require a different approach.

Palliative care for children

In two articles, "Guidelines for deciding care of critically ill or dying patients," in *Pediatrics* in 1979, and "Moral and ethical dilemma in the special care nursery" in *New Engl J Med* in 1973, R. S. Duff classified cases as A, B, or C, depending on how to treat them.^{3,4)} Class A cases should be treated as much as possible, class B should be treated selectively, and in class C cases, all treatment should be withdrawn.

M. Funato, the ex-vice president of the Yodogawa Christian Hospital, stated that in cases of 13 or 18 Trisomy, the restriction of invasive treatment for prolonging life should be earnestly considered.⁵⁾ "Smith's recognizable patterns of human malformation (7th edition)" advocates for selective treatment in which cases are handled individually, taking into consideration the child's condition and parents' wishes.⁶⁾ The categories include active treatment (all treatments are administered), restrictive treatment (certain treatments such as cardiac surgery and hemodialysis are held back), relief treatment

(further treatment is not performed, but continued treatment and nonmedical care are administered), and terminal care (all treatments are suspended).⁶⁾ The ethical and medical decision-making guideline of the Yodogawa Christian Hospital also advocates selective treatment.

While medicine seeks to offer every possible treatment when a serious disease is found in a long-awaited child, clinicians must also be conscious of medicine's limitations, and to help parents love and respect a child in the womb who is found to have an incurable disease. Nurses and midwives in our prenatal team show warm sympathy toward babies born with the fate of a short life. They welcome the baby after birth, and spend quiet time with the family. When the child dies, they bid farewell to the deceased child, dressing him or her with flowers and clothes made by volunteers who themselves have lost a child.

To determine whether a disease is incurable or not, it is critical to diagnose the condition as accurately as possible and evaluate the outcome. The evaluation of the outcome may change as improvements in medicine take place. For example, in our field, myelomeningocele (MMC) is the most common malformation. In 1974, an article by J. Lorber, "Selective treatment of MMC," was published in *Pediatrics*.⁷⁾ This report is the famous Lorber's adverse criteria, which indicate that cases with high-level MMC, deformation of the spine, severe hydrocephalus, and associated anomalies are inoperable. However, as treatments have improved, these adverse criteria have very rarely been used since the 1990s. On the other hand, improvements in prenatal diagnosis have also led to an increased percentage of TOP cases in some countries. In some regions, TOP for MMC is still strictly prohibited due to religious or political reasons.⁸⁾ Intrauterine repair of MMC was the subject of a clinical trial in 2011 called the Management of Myelomeningocele Study (MOMS), which reported some improvement over repair after delivery. At our institute, we have experienced 21 cases of prenatally diagnosed MMC since 2012, 3 of which were terminated and 18 of which were treated after birth.

Funato has stated that the most important theme in pediatric medicine is the respect of the human rights and dignity of children with a poor prognosis who cannot express themselves, in considering the best treatment. Endless overtreatment can lead to excess work for neonatologists and a shortage of beds in the NICU. To treat or not to treat can depend on the medical resources available, the social-economic status of the community, and the limits of medical science. Moral dilemmas will continue

to be addressed as future medical progress yields treatments for currently untreatable diseases.

Medical neglect

Only 8 years ago, it was very difficult to recognize medical neglect, even in cases in which parents denied their child life-saving medical care. Case 4 was the second case in Japan in which surgery was performed on a medically neglected child after the suspension of parental rights and the selection of a substitute consent giver. In the first case, the substitute was a relative, but in our case, the substitute was a non-relative. Another difference is that the baby in the first case had a fatal congenital heart disease, while the baby in our case had the potential to be disabled, but no threat to life (news.a902.net/a1/2003/0810-11.html). Despite this fact, the judgment in our case was granted at unprecedented speed, and this legal case has therefore gained a lot of attention. Notably, the opinion of the review board of the ethical committee was not unanimous. Although several members agreed that the case represented medical neglect, another member worried that "If the parents believe that child's soul will be damaged, their love toward their child will diminish after surgery and lead to child abuse."

Since then, this process has been widely applied in cases where medical neglect is feared to cause a serious condition. Notably, in reality, many doctors face situations of medical neglect and refusal of surgery. Miyamoto et al. conducted a "Survey on medical neglect in the pediatric medical field in Japan" throughout Japan, which asked clinicians to report on their experience of explicit refusal by parents for medical care for a child's disease, malformation, or injury. The results revealed that 60 hospitals out of 328 had experienced active medical refusal in 2003.⁹⁾

In April 2012, the civil law was amended so that in cases where a serious impact on a child's life and health was suspected, parental rights could be suspended and a substitute consent giver selected. This amendment made the legal process in these cases faster and much less complicated than in previous cases (<http://www.gov-online.go.jp/useful/article/201203/1.html>).

Conclusion

There are always ethical dilemmas related to innovative medical technology, and workers in

clinical fields constantly have to address them. Today, almost all hospitals have their own ethical committee, to which medical ethics cases can be presented. Even within the same medical environment, neurosurgeons, pediatricians, and neonatologists have different perspectives, views, and opinions. It is also important to hear the opinions of people outside of medicine. Moreover, the medical and ethical landscape is constantly shifting, and medical opinions and laws will change over time, according to what the majority of people believe to be true. Thus, it is vital to address ethical situations until some change providing a solution occurs.

Conflicts of Interest Disclosure

The author has no conflicts of interests to disclose.

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