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Case Study

Active Euthanasia for Perinatal Osteogenesis Imperfecta; An Ethical Dilemma in a Tertiary Facility in Southwestern Nigeria: A Case Report

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Abstract

Managing a newborn with lethal congenital anomalies is challenging but handling a parent's request for doctors under oath to terminate the baby's life is another major ethical dilemma requiring cautious evaluation. We present a term male neonate who presented on the 7th day of life, with a dark-blue sclera, multiple limb deformities, long bone fractures, beaded ribs, a flattened forehead, a narrow chest, and respiratory distress. A Diagnosis of Type II Osteogenesis imperfecta was made and he was managed by a multidisciplinary team including neonatologists, geneticists/endocrinologists, orthopaedic surgeons, nurses, and medical social workers. Supplemental oxygen, intravenous fluids and antibiotics, analgesia, and bisphosphonates were offered as supportive care. The main concern was the challenges of managing a newborn with lethal OI and balancing the demand for euthanasia by the parents to end the baby's misery. In providing care, the rights of the child to life, the morals of the physician, the best interests of the baby, and the family's role in decision-making in a setting of out-of-pocket expenditures must be weighed. Following extensive multidisciplinary team meetings, it was ultimately decided to allow nature to take her course. Baby subsequently had progressive respiratory distress from pulmonary hypoplasia and died of respiratory failure on the twelfth day of life. In Conclusion, Osteogenesis imperfecta of the perinatal type is usually a lethal disease, with death often occurring within the perinatal period. The physician must, therefore, balance the parental rights, the oath of office, and the existing legal framework to avoid charges of murder or manslaughter.

Keywords
Osteogenesis Imperfecta, Euthanasia, Ethical Dilemma, End of Life, Case Report.

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Quick Response Code:

Introduction

The intentional termination of a newborn's life within the first 28 days of life is known as neonatal euthanasia.[1] In most parts of the world, newborn euthanasia is unethical due to the physician's fiduciary duty, the Hippocratic Oath, the lack of a legal framework for such a practise, and each individual's inalienable right to life.[1] Under the ethics of practise, the medical practitioner has a duty to preserve life, to treat the sick patient with respect and compassion, and to always uphold the dignity of the individual, regardless of age, race, or religion. It has been demonstrated that euthanasia is not a viable alternative to the agony and suffering of the dying process for the terminally ill or those with lethal diseases such as perinatal type Osteogenesis Imperfecta (OI).[2, 3] The proposed reason is the absence of a dignified death, in which a protective arm is transformed into a weapon of destruction, diminishing the nobility of care and love.

In stark contrast to the preceding, the Groningen Protocol (GP) was established to justify the euthanasia of critically ill neonates based on carefully selected criteria such as "unbearable suffering" and "expected quality of life " to which some have argued that the protocol was based on a flawed moral principle, with a shift of responsibility to a parent who had placed their trust in a noble profession. The defining feature of the GP protocol is that the final decision regarding "active termination of life" rests with the parents, with physicians and social workers in agreement.[1, 3] When a well-educated father discovers the GP protocol and requests that the physician uphold the GP principle in the absence of legal support, the physician is put in a precarious situation. Neonates have the least legal right to make their own health decisions. This is especially concerning when the parent's wishes conflict with the child's inalienable right to life, making it difficult for the physician to decide what to do in the face of firm moral principles and established medical ethics. We present a case report of a 7-day-old infant with perinatal osteogenesis imperfecta whose father requested active euthanasia at the University College Hospital Ibadan, as well as a literature review.

Patient and observation

Patient Information:

A term, small for gestational age (2.0kg) male neonate was delivered vaginally at 37 weeks gestation to a 35-year-old para 2 + 0 (2 Alive) mother. Pregnancy had no negative consequences. Initial obstetric ultrasound scans in the first and second trimesters revealed no femur. The most recent antenatal ultrasound at 30 weeks revealed a foetus with a large head, shortened humerus, and bowed demineralized femur. There is no family history of OI. Referred to the emergency neonatal ward from an outlying hospital due to multiple limb abnormalities and respiratory distress observed since birth. The father is a university lecturer, and the mother is a graduate.

Clinical Findings

Examination findings at the presentation revealed a small for age infant with obviously short limbs, a narrow chest, a flattened forehead, and a dark blue sclera (Figure 1), who was in respiratory distress. Musculoskeletal examination revealed disproportionately short and deformed limbs with obvious bowing of the tibia and humerus and multiple prominences (Callus formation) over all the long bones

Figure 1: Clinical picture of a baby with Osteogenesis Imperfecta

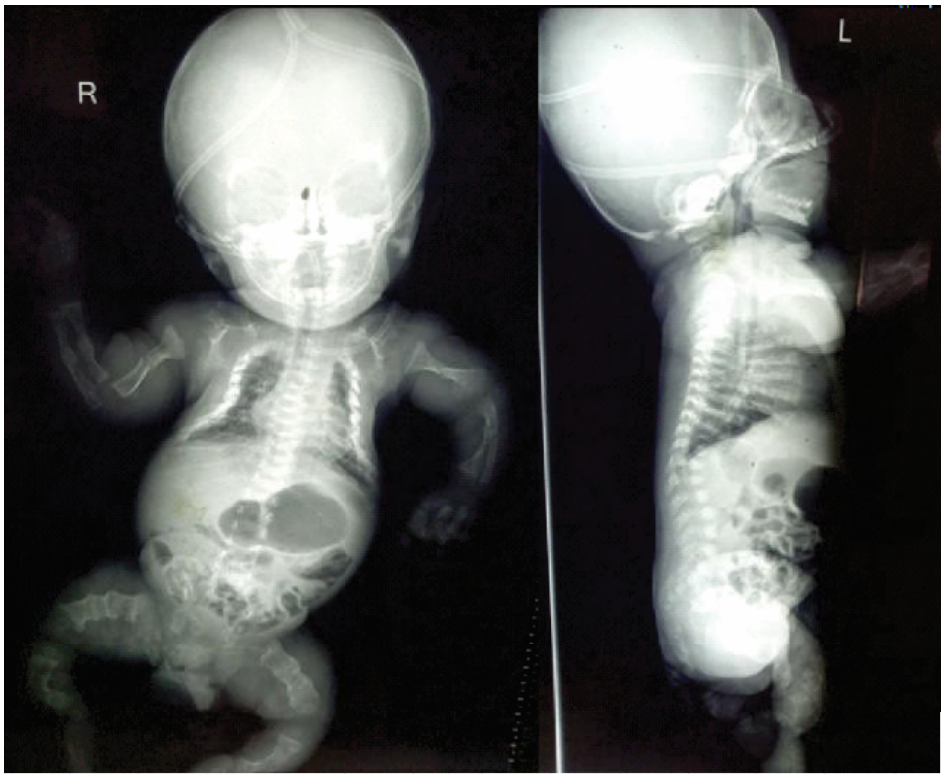

Figure 2: Babygram showing (Multiple fractures of the humerus, radius and ulnar, femur and tibia, clavicle, short and broad limbs, narrow thorax, callus formation in the long bones, beading of the ribs) in a patient with suspected Osteogenesis Imperfecta Type II

Timeline of the current episode

The details of the timeline of events are in Table 1.

Table 1: Timeline of patient management

Days of life	Events	Intervention
Conception – GA 37wks+3days	Uneventful pregnancy and delivery at a private clinic	Routine ante-natal care and delivery
Day 0 - 2	Short limbs were noticed but the baby was sucking well Parents reassured and the baby discharged	
Day 3 - 6	Parents noticed generalized tenderness of the limbs and respiratory distress	Parents continued routine care of the baby but had increasing concerns after researching about condition
Day 7 - 9	Presentation and admission with progressive worsening of respiratory distress	1. The baby was given supportive care, including supplemental oxygen, antibiotics, analgesia, and other comfort measures. The nurses make a special exception for patients with severe form osteogenesis imperfecta. 2. The Geneticist, Orthopaedic Surgeon, and Social Workers were all consulted. 3. The surgeon will provide surgery after the neonatal period. 4. Parents were offered serial counseling conference sessions with the multidisciplinary team. 5. IV Bisphosphonate was recommended. 6. The father requested euthanasia. 7. A departmental meeting was held to discuss the baby's health. Senior colleagues provided advice on how to handle the father's request, and the head of the newborn unit was notified.
Day 10 - 11	Marked clinical deterioration with recurrent apnoea	As the infant's condition worsened, respiratory support was administered.
Day 12	Developed respiratory failure	

Diagnostic assessment

Laboratory work-up at admission included packed cell volume of 58% (51± 5), complete blood count with WBC of 12,640cells/µL with differentials (neutrophils- 53.3%, lymphocyte-34.8%, monocyte-11.5%), platelet count of 444,000 cell/µl. Blood culture at admission was sterile, CRP- 6.0mg/L. Electrolytes, urea, and creatinine results showed Sodium- 147mmol/l, potassium- 4.4mmol/l, chloride-115mmol/l, bicarbonate of 16mmol/l, urea- 64mg/dl, creatinine- 0.4mg/dl, calcium- 9.0mg/dl, phosphate-4.3mg/dl. Babygram showed Multiple fractures of the demineralized humerus, radius and ulnar, femur and tibia, clavicle, short and broad limbs, narrow thorax, callus formation in the long bones, and beading of the ribs.

Diagnosis

Osteogenesis imperfecta type II was confidently diagnosed based on postnatal examination and bone radiographic findings. Genetic testing for mutations in COL1A1 and COL1A2 could not be made before Childs' demise.

Therapeutic interventions:

The neonatologist, geneticist/endocrinologist, orthopaedics, nursing, and medical social workers were all involved in the care. The patient was given respiratory support. In a high-dependency unit, he received extra gentle nursing care due to his fragile bones. He was given intravenous fluids with amino acid supplementation, as well as paracetamol, and was offered bisphosphonates. A multidisciplinary team counseled the parents on the patient's condition and prognosis on multiple occasions. On the tenth day, both parents requested euthanasia for the infant. On the contrary, the managing physician adhered to the medical ethics and the applicable law. The patient's management was continued, but he was offered a ceiling of care. Respiratory distress worsened, and the child experienced multiple apnoeic episodes during admission, necessitating ventilation beginning on the 10th day of life. On the 12th day of his life, he developed respiratory failure and suffered a terminal cardiopulmonary arrest.

Discussion

As advanced diagnostic modalities and intensive care become more widely available in low and middle-income countries, so will access to these healthcare services and interventions. More critically ill infants are surviving in low-income countries, where their diagnoses may be missed and they would not have survived otherwise.[4] As a result of these advances, there will be challenges, including ethical concerns about whether the physician should continue patient management or limit intervention to a ceiling of care to preserve limited resources, or continue care in futility.[4] Furthermore, the increased availability of critical medical information to caregivers/parents makes communication between doctors and caregivers/parents about critically ill patients more effective, but at a cost. Parents' access to prognostic indicators for various diseases, despite their incompleteness and lack of supporting evidence, may lead to the formation of erroneous conclusions against existing evidence well known to the physician. The onus is on the managing physician to ensure that the ethical principles of autonomy, beneficence, non-maleficence, and justice are not violated in decision making.

Our experience with a 7-day-old baby suffering from OI, a rare genetic disorder occurring in 1 in 10,000-20,000 births highlights the aforementioned circumstances.[5] Osteogenesis imperfecta (OI), also known as brittle bone disease, is a genetic disorder that manifests as a spectrum disorder ranging from mild type I to type VII variant, which is most common in Australians with a milder phenotypic presentation, to the most lethal type II, also known as the perinatal type [2] The diagnostic triad of blue eyes, short stature, and multiple fractures caused by brittle bones serves as the basis for diagnosis. The most severe form of

perinatal osteogenesis imperfecta is frequently diagnosed in utero by chorionic villus sampling in high-risk pregnancies as early as the late first trimester (11th week) to the early second trimester. On the samples, histology and genetic studies are conducted. In sporadic cases in which there is no family history of the disease, a routine antenatal ultrasound or anomaly scan performed between 18 and 21 weeks of gestation may provide the initial diagnosis.[6] On the basis of the findings, parents are counseled about the disease and given prognostic indices and potential differential diagnoses. Also discussed are the potential short- and long-term effects of the disease. This affords the parents ample time to make important decisions. The managing physician is responsible for informing the parents of the diagnosis and providing available support, management plans, and a prognosis if the antennal diagnosis window is missed or the pregnancy is carried to term. Unfortunately, when a postnatal diagnosis is made, the parents/caregivers are typically in an emotional state and may not be able to make an informed decision. Such a family may require substantial emotional support to cope with the reality of their child's clinical condition. In our case, the parents were not notified until they were referred to our facility for specialised care.

Following a series of counseling by the multidisciplinary team and having allowed the parents time to ruminate over the information provided. The fathers allude to further reading on care of end-of-life care. The physician was approached to voluntarily terminate the neonate's life to relieve him of the ongoing misery. This is particularly concerning given the low value placed on newborn lives in low-income countries. These infants are susceptible to maltreatment, particularly those with potentially fatal congenital anomalies and those with the potential for negative long-term outcomes.[7]

Providers of health care must possess pertinent skills and knowledge, as well as be familiar with the ethical and legal expectations that result from standard practices.[8] The inalienable right to life is enshrined in Chapter IV of the 1999 constitution of the Federal Republic of Nigeria (as amended by Section 33).[9] Any attempt to revoke this right is unlawful and punishable by law. Nigeria's constitution contains no provision for euthanasia, a procedure in which a terminally ill, severely ill, and competent patient, or his legal surrogate, gives consent to end his or her life. If all legal requirements are met, there is an exception to this rule: the judicial process has the authority to decide whether or not to impose the death penalty. Any physician who commits this act could be charged with murder or manslaughter.

Except in Sweden, where the Groningen Protocol provides an exception based on specific eligibility requirements, most countries do not offer the option of neonatal euthanasia. In a review of published literature on GP, Alves Daniela and Costa concluded that the arguments against the GP were far superior to those in support.[10] Although the GP was presumably founded on the four pillars of medical ethics (respect for autonomy, beneficence, non-maleficence, and justice), they trust it was designed primarily to target the deliberate termination of a newborn's life without fear of criminal prosecution.[10]

When a request for euthanasia is made in a low- and middle-income country (LMIC), the physician's initial options may include but are not limited to, contacting senior colleagues who have either experience with the issues at hand or have been certified by the country's legal body to administer law in addition to their medical qualifications. Because the hospital is responsible for every patient's care, the Chairman Medical Advisory Committee (CMAC) of the hospital should be consulted. In addition to the CMAC, the hospital ethics board can provide advice based on medical ethics principles. As the apex of the medical practice framework in countries where it is active, the National Sovereign Council for Medical Practice provides support and legal counsel. Finally, the hospital legal department is in charge of providing legal counsel and representing the legal apparatus of the state. If necessary, they act as a liaison between the hospital's managing physician and the legal system such as the magistrate court. They interface to obtain

a legal injunction to protect the minor's rights, even if this violates the caregiver's autonomy. The oath of office instills a fundamental instinct in a physician that cannot be overstated. It should be emphasized that as all of the above interventions have been considered, the continuation of care and provision of comfort to an ill child must not be jeopardized.

On the other hand, low-income countries' legal systems must adapt to the ongoing trend of political, economic, cultural, and technological advancement. Furthermore, as new medical interventions become available, it is reasonable to expect physicians to be exposed to new legal risks that did not previously exist. It is therefore expedient that a periodic medical referendum be held in order to identify legal gaps that need to be filled, practitioners' current dilemmas, and shifts in practitioners' beliefs. As the population becomes more informed and patients have access to the global village with its diverse laws, medical professionals must be familiar with the laws governing their field. End-of-life care should be provided by a multidisciplinary team, including the spiritual head of a hospital, who will handle the spirituality and religiosity of caregivers to provide appropriate support. The importance of family-centered care cannot be overstated, as primary care providers are frequently in a vulnerable mental state.

Conclusion

Osteogenesis imperfecta of the perinatal type is usually a severe and lethal disease, with death often occurring within the perinatal period. The physician must balance the parental rights, the oath of office, existing legal framework to avoid charges of murder or manslaughter.

Patient Perspective

The caring doctors and nurses, as well as the information we found online, helped us understand how our baby's illness was passed down, how it showed up in the hospital, and what the possible outcomes were. Our baby was the result of an unplanned pregnancy, and it was born with a fatal birth defect. Seeing our baby in pain made us think about asking doctors to end the suffering. We decided to stay because the doctors emphasized the morality of our request. We decided to wait and see what happened because we believe that God knows everything and is in control. So,

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References

1. Kon JJ, Verhagen A, Kon AA. Neonatal Euthanasia and the Groningen Protocol. in *Pediatric Ethics: Theory and Practice*: Springer, 2022:291-311.
2. Ayadi ID, Hamida EB, Rebeh RB, Chaouachi S, Marrakchi Z. Perinatal lethal type II osteogenesis imperfecta: a case report. Pan Afr Med J. 2015;21:11. doi: 10.11604/pamj.2015.**21**.11.6834.
3. Wach B. Dutch protocol from Groningen (the so-called Groningen Protocol). The problem of deliberate causing of death in newborns ("neonatal euthanasia"). Analiza i Egzystencja. 2020;**49**:53-87.
4. Losonczy LI, Papali A, Kivlehan S, Calvello Hynes EJ, Calderon G, Laytin A, et al. White paper on early critical care services in low resource settings. Ann Glob Health. 2021;**87**(1):105. doi: 10.5334/aogh.3377.
5. Lim J, Grafe I, Alexander S, Lee B. Genetic causes and mechanisms of osteogenesis imperfecta. Bone. 2017;**102**:40-9.

6. Rumack C, Johnson M, Zunkel D. "Antenatal diagnosis of osteogenesis imperfecta Clinical Diagnosis with Ultrasound. 1981;**8**:210.
7. Alao MA, Ibrahim OR, Ogunbosi BO, Nna EO, Olapegba PO. Effectiveness of Faith-Based Interventions on the Rate of Discharged Against Medical Advice in Tertiary Newborn Units in Nigeria: A Protocol for an Open Label Randomized Control Trial. Front Public Health. 2021. 2022.1;9:788383.
8. Monsudi KF, Oladele TO, Nasir AA, Ayanniyi AA. Medical ethics in sub-Sahara Africa: closing the gaps. Afr Health Sci. 2015;**15**(2):673-81
9. Nigeria's Constitution of 1999, available at https://www.constituteproject.org/constitution/Nigeria_1999.pdf, accessed on 23/11/2022.
10. Alves D, Costa ED. The Groningen Protocol for neonatal euthanasia: Our perspective. Nascer e Crescer - Birth and Growth Medical Journal. 2019;28: 185-190.