The Ethics of Lethal Congenital Malformations. Is Palliative Therapy Possible?

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Aim: This study aims to show that palliative care can be offered to babies born with lethal malformations according to the Islamic Code of Ethics, which takes into account the well-being of the mother and her fetus. Methods: For our study, a lethal congenital malformation is defined as a condition that invariably leads to death in utero or the newborn period regardless of attempted supportive care. The cases were identified during weekly prenatal meetings, which discuss abnormal fetal ultrasound findings and genetic testing to provide plans for their management. In addition, cases that were not diagnosed antenatally but diagnosed at birth were also included. The management plans for those cases with lethal malformations were discussed before birth, and the families were counseled, and their consent was taken for nonmonitoring of the fetus during pregnancy and noninterference with the newborn baby except for palliative care. Families who requested full support during pregnancy and delivery were given that option. Results: Over a period of 16 years from 2001 to 2016, a total number of 92705 babies were born in our hospital. Antenatal ultrasounds were performed by the feto-maternal physician team. There were 1563 abnormal fetal ultrasounds (1.7%) with significant various organs malformations. Of these abnormal ultrasounds, 384 were diagnosed as possible lethal congenital malformations (24.6%). The remaining abnormal fetal ultrasounds were compatible with life and were managed according to the standard clinical practices. Fetuses diagnosed with lethal malformations were discussed at length and parents were informed about the lethal outcome and the plan of nonmonitoring of the mother at delivery and avoidance of cesarean section except for maternal indication and eventual palliative care for the live newborn. Parents’ consent was obtained, and the decision and the plan of the management are documented clearly in the mother’s medical record file. With this management plan, we were able to offer palliative care to the cases of lethal congenital malformations who eventually expired within a short time. Conclusion: Within the guidance of the Islamic Jurisprudence (Figh), it is possible to manage cases of congenital malformations, which are considered incompatible with life palliatively and avoid unnecessary cesarean sections in the mother. This practice has resulted in better utilization of the beds in our busy neonatal intensive care unit. Keywords: Lethal malformations, neonatal intensive care unit, newborns, palliative therapy, prenatal

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This is particularly true for the obstetric and neonatal services where the well-being of the mother, her fetus, and the newborn is the focus of that care. In the days, when we did not have specific information and prenatal diagnosis of malformation of the fetus, the newborn was given full respiratory support and intensive care for a prolonged period. Babies with lethal malformations were ventilated because the definitive diagnosis was delayed.\[1\] Examples of these include pulmonary hypoplasia in Potter cases, thanatophoric dysplasia, trisomy 18, and trisomy 13. However, with technological advances in the field of antenatal sonography and genetic testing, it is now possible to diagnose a large number of fetal malformations and genetic disorders with certainty, and this is particularly true for some lethal congenital malformations. In making decisions of the management of the malformed fetus, the Muslim doctor is guided by the Islamic Code of Ethics which emphasizes the importance of the well-being of the mother and her fetus and later her newborn baby.\[2,3\]

**Aim of the study**

The aim is to show that palliative care can be offered to babies born with lethal congenital malformations without undue sufferings and that with accurate diagnoses these babies can be managed with minimal intervention for both mother and baby.

**Design**

A prospective observational study carried out jointly by the Obstetric and Neonatal Departments at Security Forces Hospital where all pregnancies were monitored antenatally, and abnormal ultrasounds and test results were discussed weekly and plans made for the time and mode of delivery of the mother and after care of the newborn.

**METHODS**

This prospective study was conducted at Security Forces Hospital in Riyadh between the years 2001 and 2016 and was approved by the Hospital Research and Ethical Committees. The study was aimed toward quality improvement in the care, which is provided at the perinatal level. Over the years, improvements in ultrasound techniques and their interpretations and genetic testing during pregnancy have enabled the physicians to plan in advance for the delivery and care of the at-risk mother and her fetus. Weekly prenatal meetings were held to discuss abnormal antenatal findings to provide management plans for the various disorders encountered. A record of the cases discussed was kept at hand in the emergency room, delivery room, and neonatal intensive care unit (NICU) together with the management plans. Other cases where there was a family history of a medical disorders resulting in fetal loss or neonatal death were also recorded. Cases were labeled as lethal malformations when the diagnosis and/or prognosis are certain to end in death. The management plan endorsed, depended on the opinion expressed by the Council of Islamic Jurisprudence (Figh)\[4\] and a number of decrees (Fatwas) issued by the Permanent Committee for Scholarly Research and Ifta in Saudi Arabia in (Fatwa number 12086, 12762, and decree 190)\[5-7\] which clearly indicated that resuscitation or life support could be withheld if the patient is suffering from an incurable illness that is not responding to treatment and their death, is certain.

The management plans for antenatally diagnosed cases of lethal congenital malformations were thus discussed with the relevant families, and the outcome explained clearly. For these cases, parents’ consent was obtained before delivery, and a decision not to monitor the mother during labor is documented clearly in the mother record together with the decision to offer the baby palliative care consisting of feeding, warmth and pain relief but no resuscitation. However, for families who opted not consent to our plan of nonmonitoring during labor full support was given, including cesarean section and resuscitation of the baby.\[1\]

All cases of lethal congenital malformations were registered whether live born or stillborn and their diagnoses confirmed. The exact time of death was recorded for each case to define the average survival time for each disorder.

**RESULTS**

Over 16 years’ period, a total number of 92705 babies were born. Antenatal ultrasounds performed by the feto-maternal team identified 1563 fetuses with various malformations (1.7%). Out of the 1563, there were 384 (24.6%) ultrasounds with lethal congenital malformations. The remainders of the abnormal antenatal ultrasounds were compatible with life and were managed according to the standard practice in each case. The cases discussed because of family history of genetic disorder and/or unexplained neonatal death resulted in 63 normal newborns and 17 babies with genetic disorders. The distribution among these genetic cases consisted of 8 cases of neurological genetic disorders, 5 cases of inborn errors of metabolism associated with primary or secondary lactic acidemia, 2 cases of genetic neuromuscular disorders, and 2 cases of immune deficiency. None of these were considered lethal and were given full care and support.

The distributions of different groups of disorders among lethal malformations are shown in Table 1. The majority of lethal malformations were due to
Potter disorders (89 cases) making 23.2% of the lethal malformations. About 70% of the Potter cases were due to polycystic kidney disease (Potter Sequence) and 30% were cases of renal agenesis (Potter Syndrome). All the Potter cases were diagnosed antenatally and were associated with severe oligohydramnios and pulmonary hypoplasia.

There were 67 (17.4%) cases of severe multiple anomalies which were considered lethal.

The lethal central nervous system malformations were common constituting 16.9% of the total lethal malformations amounting to 65 cases distributed between 26 cases of anencephaly, 18 cases of hydrocephalus with associated severe multiple anomalies, 13 cases of Meckel-Gruber syndrome, and 8 cases of Walker–Warburg syndrome.

Lethal chromosomal disorders were 47 cases (12.2%) distributed equally between trisomy 18 and trisomy 13.

Congenital lethal cardiac malformations accounted to 37 cases (9.6%) presenting as hypoplastic left heart syndrome.

Although the overall survival of isolated cases of congenital diaphragmatic hernia has improved, there are still cases that are associated with other congenital anomalies or chromosomal aberrations that render them lethal. As such 25 cases of diaphragmatic hernia were encountered and ended lethally.

There were 21 cases of lethal osteochondrodysplasia consisting mainly of cases of thanatophoric dysplasia. Other cases included achondrogenesis type 1 and dysplasia with severe multiple anomalies.

Lethal nonimmune hydrops occurred in 10 cases and were associated with pulmonary hypoplasia or neurometabolic disorders. The remaining cases were due to tracheal atresia and hemophagocytic lymphohistiocytosis (HLH).

The survival of the babies with lethal congenital malformations varied from few hours to 30 days [Table 2]. In general, babies with Potter disorder, skeletal dysplasia, Meckel-Gruber syndrome, and tracheal atresia survived for 1–6 h while babies with anencephaly and nonimmune hydrops survived for 72 h. Babies with lethal chromosomal disorders, hypoplastic left heart syndrome, and Walker–Warburg syndrome survived for between 7 and 14 days. The longest survival occurred in cases of severe multiple congenital anomalies and hydrocephalus with multiple anomalies, lactic acidemias, and HLH.

In our cases of lethal malformations, there was a strong association with oligohydramnios and polyhydramnios.

Oligohydramnios was associated with 173 (45%) cases of lethal malformations while polyhydramnios was associated with 77 (20%) cases. The majority of cases of lethal malformations associated with oligohydramnios were due to Potter disorder (83%), whereas those associated with polyhydramnios were mainly due to severe multiple anomalies and trisomies.

**DISCUSSION**

In our NICU, lethal congenital malformations are the most common cause of neonatal mortality amounting to >50% of the neonatal deaths. In the past, when a definitive diagnosis of lethal malformations could not be made antenatally or soon after birth, a large number of babies were given full respiratory support and intensive care.\(^{(1)}\) Because these babies survived for longer periods, the NICU had to cope with overcrowding,
repeated nosocomial infections with the emergence of multidrug-resistant bacteria, prolonged ventilation, and poor utilization of resources. With the improvement of diagnostic methods and obstetric ultrasound technology, it became evident that many of the lethal disorders can be diagnosed antenatally.[1] The next step for us was to define what a lethal malformation is and whether it can be managed palliatively according to the Islamic Code of Ethics.

The definition we adopted for a lethal malformation was any condition that invariably leads to death in utero or the newborn period regardless of attempted supportive care.[8] According to this definition, lethal malformations are classified into three groups on the basis of certainty of the diagnosis and certainty of the prognosis:[9]

a. A group where the diagnosis is certain and prognosis is certain to end in death. This group can be diagnosed antenatally by ultrasound or amniocentesis for chromosomal and genetic disorders and can be managed palliatively without intensive intervention. Examples include trisomy 13, trisomy 18, triploidy, anencephaly, holoprosencephaly, aracdia, and Potter cases associated with renal agenesis or polycystic kidney disease.

b. A group where definitive diagnosis is not attainable but lethal prognosis is certain. Examples include fetuses with early oligohydramnios and pulmonary hypoplasia, severe congenital hydrocephalus with absent or minimal brain growth and certain forms of skeletal dysplasia such as thanatophoric dysplasia. When the prognosis is a certainty palliative care can be offered as a standard of care.

c. Some conditions with a definitive diagnosis have a prognosis, which may not be clearly terminal. Examples include hypoplastic left heart syndrome, multiple severe anomalies, severe cases of neurodegenerative disorders, and some forms of errors of metabolism that are lethal even in the presence of treatment.

For the purpose of management of these cases, palliative care is defined as the active, total approach to care, embracing physical, emotional, social, and spiritual elements.[9] It focuses on what is the best option for the fetus or newborn with a known lethal anomaly, support for the family and include management of distressing symptoms and provision of support and care through death and bereavement.[9] In the Muslim Society, the spiritual element plays a major role when palliative management decision is considered. For this reason, we had to seek the advice and opinion of the Islamic Jurisprudence Scholars since the issue of sacredness of human life is one of the fundamentals in Islamic teaching. The Islamic Code of Ethics considers that human life is sacred and should not be willfully taken except on the indications specified in Islamic Jurisprudence all of which are outside the domain of medical profession.[2,3] The sanctity of human life covers all its stages, including intrauterine life of the embryo and fetus. This was clearly stated in Quran in Sura 5 verse 32 “On that account we decreed for the Children of Israel that whoever kill a human soul for other than manslaughter or corruption in the land, it shall be as if he killed all mankind, and who-so-ever saves the life of one, shall be as if he saved the life of mankind.” Thus, the doctor shall not by any means compromise the life of any patient except for the absolute necessity recognized by Islamic Jurisprudence.[4] It is noteworthy that Islamic law empowers Muslim Courts and Scholars to determine each case of terminal illness. Decisions are deducted from Quran (The Holly Book), Sunna (Prophet Instructions), tradition and cultural understanding of nature of life in the community. Although it may be possible to recognize the burden of terminal illness on the family, there is no justification in Islam to end human life merely because of the family and patient sufferings.[3] In his defense of life, however, the doctor is well advised to realize his limits and not transgress it. If it is scientifically certain that life cannot be sustained, then it is futile to diligently keep on the vegetative state of the patient by heroic means of animation or preserve him by other artificial methods.[5] This was supported by a decree (Fatwa) number 12086 issued in 1984 by the Permanent Committee for Scholarly Research and Ifta in Saudi Arabia.[5]

Having clarified the situation for palliative management of lethal congenital malformations from an Islamic point of view, we started to apply this to our babies. The pregnancies, which are followed in our institution, are all high-risk pregnancies, which are screened for anomalies by the experienced feto-maternal team. The abnormal fetal ultrasounds and genetic testing are discussed in a weekly joint prenatal meeting between the obstetric and neonatal departments and management plans are made for the delivery of the mother and the care of her newborn. We conducted this study to prove that what we are practicing does not violate the ethics of Islam toward human life.

All the cases in our series satisfied our definition of lethal malformation, which meant that the decision was not to monitor the mothers during labor and that their lethally malformed babies were to be given palliative care. Consent of the parents was obtained before delivery of the baby. The cases where the family did not consent to palliative care were given full support to the mother and her baby.[1] Palliative care consisted of providing...
comfort, oxygen, nutritional support, and paracetamol for pain relief. Babies were kept warm and turned from time to time, and nurses ensured changes of the diaper frequently and were dressed and kept in respectable appearance. Parents were given support in NICU and were encouraged to interact with their babies.\[^9\]

The types of lethal congenital malformations that we encountered are shown in Table 1. We have shown that the majority of babies who were delivered survived for few hours to few days. The longest survival was 30 days for those babies with severe multiple anomalies [Table 2].

An important factor, which helped in the diagnosis of a lethal malformation, was the presence during the pregnancy of oligohydramnios or polyhydramnios.\[^9\]

These two findings were associated with 65% of all cases of lethal malformations. Oligohydramnios tended to occur more frequently with cases of lethal malformations particularly when it occurs early in pregnancy resulting in severe pulmonary hypoplasia. Table 3 shows the types of lethal malformations which were associated with oligohydramnios and polyhydramnios. The group of lethal malformations that were identified in our series represents only few of the lethal malformations which have been reported in the literature.\[^9\]

None of the genetic disorders were labeled as lethal, and the pregnancy was allowed to proceed as normal and the newborns were given support and care after confirmatory tests were available.

Because we have been practicing palliative management for lethal congenital malformations, it is legitimate to question whether some of these cases were offered abortion or termination of pregnancy. The situation for abortion was clarified by the Islamic Jurisprudence (Figh) Council of the World Islamic League in its decree (Fatwa) number 4 in its 12\(^{th}\) session held on February 1990.\[^4\] The Fatwa states that for abortion to be allowed certain criteria must be met:

1. Committee of specialized competent physicians
2. Fetus is grossly malformed

3. Malformation should be diagnosed as untreatable, serious and unmanageable or the life of the malformed fetus would be a misfortune for itself and the family.

The time for abortion to be performed should be before the 120\(^{th}\) day of conception computed from the date of fertilization and not from the date of the last menstrual period. Abortion after 120 days of conception is allowed if the pregnant mother suffers from a disorder that endangers her life.\[^2,3\]

In our cases of lethal malformations, abortion could not be considered because the diagnosis could not be established early in the first trimester.

The termination of pregnancy for lethal malformation was discussed in the 12\(^{th}\) session of Muslim World League of Jurists in Makkah in February 1990.\[^4\]

Although some of the scholars believe that termination could be possible before ensoulment, it remains a controversial issue and many scholars believe it should not be allowed.

We think that our palliative management of pregnancies with lethal malformations has helped us in several ways to provide better care for the benefit of the family, society, and hospital quality management. This has been reflected in our practice as follows:

1. Avoidance of unnecessary cesarean section to the mother
2. Early notification of the family of any fetal anomaly and hence their involvement in decision-making
3. Improved bed utilization in NICU by withholding respiratory support and intervention in the affected baby
4. Registry of families with recurrent genetic disorders and provision of genetic counseling
5. Identification of the relevant genes associated with these malformations and hence the provision of preimplantation genetic diagnosis to the family where the gene was identified.
6. Knowledge of malformations in certain groups and tribes because consanguineous marriages are common in the Middle Eastern societies.

### Table 3: Lethal Malformations Associated with Oligohydramnios and Polyhydramnios

<table>
<thead>
<tr>
<th>Oligohydramnios</th>
<th>Polyhydramnios</th>
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<tr>
<td>Potter syndrome/sequence</td>
<td>Severe multiple anomalies</td>
</tr>
<tr>
<td>Meckel-Gruber syndrome</td>
<td>Anencephaly</td>
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<tr>
<td>Trisomy 13 and trisomy 18</td>
<td>Trisomy 13 and trisomy 18</td>
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<td>Severe multiple anomalies</td>
<td>Skeletal dysplasia</td>
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<td>Hydrocephaly with multiple anomalies</td>
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application of supportive measures to the newborn whose condition is known to be lethal. At the same time, the practice has resulted in better utilization of resources in the NICU.

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Conflicts of interest
There are no conflicts of interest.

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