Otocephaly Complex: Case Report, Literature Review, and Ethical Considerations

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Otocephaly complex is a rare and usually lethal syndrome characterized by a set of malformations consisting of microstomia, mandibular hypoplasia/agnathia, and ventromedial malposition of the ears. Those cases that have been diagnosed prenatally have used an ex utero intrapartum treatment procedure to establish a definitive airway. However, prenatal diagnosis continues to be challenging, primarily because of poor diagnostic sensitivity associated with ultrasonography. We present a case of a newborn with an unanticipated otocephaly complex requiring emergent airway management. In this report, we discuss the medical and ethical issues related to the care of a newborn with this frequently fatal condition. (A&A Case Reports. 2016;7:44–8.)

Otocephaly complex is a rare congenital disease that presents with microstomia, mandibular hypoplasia or agnathia, and ventromedial malposition of the ears.[1] These findings lay on a spectrum that may also include aglossia, cleft palate or lip, cyclopia, holoprosencephaly, and situs inversus.[2] The anatomic anomalies are secondary to failed mandibulofacial development by mesenchymal cells of the first pharyngeal arch.[3] Most sources implicate both genetic and teratogenic factors in the development of these malformations.[4] The incidence is <1 in 70,000 births, and prenatal diagnosis remains a challenge.[5,6] Only 7 patients have survived infancy.[7] In general, this condition is considered lethal because of the high prevalence of respiratory failure.

Tracheal intubation of these neonates is characteristically impossible because of stomal choanal atresia, hypopharyngeal stenosis, or upper airway obstruction.[8] Several cases, which were diagnosed prenatally, have used an ex utero intrapartum treatment procedure. This allows for partial delivery of the fetus and continued placental function while an airway is established.[7,9–11] Of the long-term survivors, all have required tracheostomy, as well as a feeding or gastrostomy tube, and the majority received reconstructive mandibular surgery.[12–15]

We present a case of a neonate delivered with an unanticipated otocephaly complex who required emergent airway management. In this report, we discuss the ethical quandaries associated with the care of a newborn presenting with both a confounding and a probable lethal prognosis.

The patient's family has reviewed this case report. Written permission for the authors to publish the report was obtained.

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CASE REPORT

The patient's mother was a 20-year-old G4P1111 (71.4 kg, height 1.67 m) who presented at 31 weeks of gestation with preterm labor. The mother had a history of hypothyroidism, which was treated with levothyroxine 50 μg daily, as well as major depression with a prior suicide attempt. She denied any family history of congenital anomalies, stillbirth, miscarriage, or consanguinity. The mother's obstetric history included 1 preterm delivery, 1 term delivery, and an ectopic pregnancy for which she underwent a laparoscopic salpingectomy.

This pregnancy had been complicated by *Escherichia coli* pyelonephritis for which the mother had received nitrofurantoin suppression therapy until 5 months of gestation. Her quad screen (α-fetoprotein, human chorionic gonadotropin, estriol, inhibin-A) performed at 28 weeks of gestation was normal.

On initial examination of the mother, her cervix had dilated to 6 cm and a cephalic presentation was noted. The fetal heart tracing was classified as a category 1 with active movement. A bedside ultrasound identified the fetus with a vertex presentation, an anterior placenta, grossly normal amniotic fluid level, and an estimated weight of 1650 g. The fetal heart rate was 130 bpm with both moderate variability and accelerations. No decelerations were noted. The mother was admitted with the diagnosis of preterm labor and pretreated with betamethasone, magnesium sulfate, and penicillin G. Seven hours later, her cervix was dilated to 9 cm and she was transferred to the labor and delivery operating room for imminent delivery.

The neonate was delivered vaginally. She was not crying and had no respiratory effort. Her APGAR scores at 1, 5, and 10 minutes were 2, 2, and 1, respectively.[16] The neonatal intensivist evaluated the patient and noted congenital malformations of the head with microstomia, mandibular hypoplasia, and low-set ears. Bag-mask ventilation was immediately initiated by a single provider. At this point, the on-call anesthesiologists were summoned. Upon arrival of the anesthesia team, the neonate was noted to be cyanotic with an oxygen saturation of approximately 70%. A 2-person technique was subsequently used for bag-mask ventilation. Air was found to be emanating from both the patient's ears, which were on her neck. Successful mask ventilation was achieved by covering her ear canals

Figure 1. Lateral view of the neonate demonstrating marked mandibular hypoplasia.

Figure 2. Frontal view of the neonate demonstrating microstomia and synotia. Permission was granted by parents to use the photos in this case report.

bilaterally with her earlobes while extending her neck. This maneuver maintained her oxygen saturation between 90% and 100%. The trauma surgery team then arrived and performed an emergent tracheostomy using a 3.5-mm tracheal tube while 2-person bag-mask ventilation continued. She subsequently had a cardiopulmonary arrest within several minutes. At that point, the neonatologist made the decision not to perform chest compressions or to administer medications based on the neonatologist's clinical judgment that the efforts were futile. Following the mother's wishes, the parents were given time to hold their daughter. Once they were ready, the patient was transferred to the neonatal intensive care unit where comfort measures were initiated. She soon died, and bereavement support was provided to the family.

Her autopsy report revealed marked mandibular hypoplasia (Fig. 1), microstomia (Fig. 2), oropharyngeal hypoplasia, a probe-patent communication between the oropharynx and the trachea, and synotia. These were consistent with otocephaly complex. The communication opened from the oropharynx into a narrowed upper trachea. Below the tracheostomy site, the trachea was of normal caliber with no appreciable obstructions. The ears were low-set and came together under her malformed mandible.

The pathology report revealed no other abnormalities. The body was that of a well-developed well-nourished female neonate weighing approximately 1435 g and measuring 35 cm. A microscopic examination revealed no significant histopathology.

DISCUSSION

Medical Knowledge

Fetal exposure to teratogens has been implicated in numerous case reports of otocephaly. Abnormal facial development has been reported following in utero exposure to smoking, alcohol, radiation, streptonigrin antibiotics, trypan blue, theophylline, beclomethasone, salicylates, amidopyrine, mycophenolate, and phenytoin.[8,17–22] The neonate in this case had a nitrofurantoin exposure during 1 to 5 months of gestation because of recurrent episodes of maternal urinary tract infections and pyelonephritis. In addition, levothyroxine was administered to the mother throughout the gestational period. However, there is no current association between the maternal use of these medications and the subsequent occurrence of otocephaly complex.

Prenatal diagnosis of otocephaly complex remains difficult despite recent advances in prenatal imaging. Several cases have made the use of 3D or 4D ultrasonography, as well as fetal magnetic resonance imaging to diagnose this syndrome.[23,24] Many were found incidentally through careful examination because of other existing pathology.[25,26]

This mother had ultrasonography at 10 weeks of gestation and during labor. Both failed to reveal craniofacial abnormalities. Ultrasonography of fetal facial abnormalities remains challenging. Typically, most craniofacial abnormalities are detected between 16 and 18 weeks of gestation. However, some have been detected as early as 10 to 12 weeks of gestation.[27] Polyhydramnios, which sometimes suggests the presence of fetal malformations, was absent on this patient's ultrasound examination. Failure to diagnose

these abnormalities with ultrasound meant that an ex utero intrapartum treatment procedure, with a surgeon available for immediate tracheostomy, could not have been planned.

Possible factors that may have led to the neonate's cardiopulmonary arrest included complications associated with the tracheostomy: false passage formation, improper tracheal tube placement, or accidental decannulation. Any of these may exacerbate the preexisting respiratory insufficiency associated with the neonate's underdeveloped lungs, subsequently leading to hypoxemia and bradycardia. Premature infants with numerous comorbidities are at especially high risk of tracheostomy-associated complications unrelated to the surgical technique.[28–30] Evidence of tracheostomy complications may not be evident on autopsy because antemortem tube position may be impossible to assess, mucoid material may have already been suctioned, and meticulous neck dissection may be required to exclude false passages and fistulae formation.[31] These complications have been linked to lower birth weights, congenital defects, and higher ventilatory pressure requirements.[28]

Future research to improve these neonates' chances of survival may involve maintenance of uteroplacental circulation after vaginal delivery. Delayed cord clamping and continued placental support, such as gently wrapping the umbilical cord in a warm wet towel, may afford the surgeon more time to perform a tracheostomy. However, this would require a well-coordinated surgical team effort. The mean duration on uteroplacental bypass (from uterine incision to umbilical cord clamping) has been shown to be 30.3 ± 14 minutes, but the optimal time for cord clamping has yet to be established.[32]

Placental support techniques, such as nitroglycerin or tocolytic administration and avoidance of uterotonic agents, can delay placental separation from the uterine wall and their use should be further explored. Despite the risks of neonatal overtransfusion and maternal infection or postpartum hemorrhage, active management of the third stage of labor has been shown to reduce the risk of hemorrhage to <1000 mL.[32] In these cases, the benefits of maintaining uteroplacental circulation would therefore appear to outweigh the risks to the mother.

Ethical Views

Within 1 hour after birth, this neonate developed cardiopulmonary distress, after an awake tracheostomy, and no further aggressive measures were pursued. The ethical issues presented herein are 2-fold. First, should aggressive resuscitation be performed or should comfort measures be provided for a neonate with otocephaly complex? Second, once aggressive resuscitation is chosen, can further interventions be withheld and care be withdrawn, if these interventions are deemed futile?

Ideally, physicians should have an antenatal discussion with the parents regarding the expected prognosis of the neonate, the neonate's quality of life, the benefits and burdens of treatment, and the situations in which treatment would be considered futile. Futile care, which is care that, in the best judgment of physicians, will not reasonably benefit the patient, is not an ethical obligation.[33] After discussion with the family, a decision regarding the anticipated care of the neonate should be made by all parties involved. In challenging situations, an ethics consultation can be requested.

This case was complicated by the altogether unanticipated presentation of otocephaly. There was no opportunity for appropriate preparation by the various medical teams or for discussion with the family to ensure an informed decision. In addition, both parents spoke only Spanish and no interpreter was immediately available. The inability to immediately communicate the details of the case to the family exacerbated this challenging situation. Although ventilation via bag-mask technique was adequate, it was unclear for how long the neonate could maintain saturation while a Spanish interpreter was located for the parents. They were thus unable to make truly informed decisions on care for their newborn. Without parental input, the neonatologists made treatment decisions based on clinical judgment alone.

Although the majority of neonates with otocephaly complex die within the first few hours of life, there is no prognostic algorithm.[8] It is conceivable that this neonate could have survived until adulthood because the autopsy report was unremarkable other than for otocephaly. However, this was unlikely given the difficulties associated with achieving adequate ventilation of this patient. There are few reports of nonlethal cases of otocephaly complex.[7,13–15] Recently, 4 patients with agnathia-otocephaly complex, who underwent successful facial reconstructive surgery, have been reported.[34]

Our patient was apneic at birth with congenital defects of unknown extent. The initial decision to begin resuscitation was made by the neonatologist, acting as an advocate for the patient's life based on the bioethical principle of beneficence.[35]

When there is unanticipated delivery of a neonate with otocephaly complex, and the neonate's medical prognosis is unknown, the ethical course of action is clearly to sustain life. However, once the decision has been made to sustain life, which in this case was to establish an airway, further decisions for continuing life-sustaining treatment should be predicated on frequent clinical evaluations. If possible, the family should be updated on the neonate's clinical status and prognosis.

The second ethical issue arose when the patient developed cardiac arrest after her tracheostomy. At our institution, the medical responsibility of the neonate transfers, at birth, to the neonatologist. The neonatologists decide, with the parents, if further aggressive resuscitation efforts should be pursued or care withdrawn. In institutions or situations where there is no neonatologist available, the responsibility for this discussion with the parents may include the pediatrician, anesthesiologist, or obstetrician.

Because of the acute nature of this case, the neonatal intensivist made the decision to begin initial resuscitation and establish a secured airway. Establishment of a secured airway allows the medical teams to further examine the neonate, have a more informed discussion with the family regarding the neonate's prognosis, and provide the family with more time to determine wishes. However, once the neonate arrested within minutes after the tracheostomy, all aggressive measures were discontinued. No chest compressions or medications were administered, and the tracheostomy was decannulated. This unilateral decision was made

by the neonatologist based on her clinical judgment that the efforts were futile. There was insufficient time to discuss the neonate's rapidly deteriorating clinical picture with her parents.

Because of improvements in medical knowledge and technology, physicians may find themselves able to sustain or extend life in neonates whose chances of surviving are minimal.[36] It must be determined by the clinician if further aggressive interventions, such as the initiation of cardiopulmonary resuscitation, would be futile.[37] The determination of futility is ultimately a value judgment.[38]

The ethical decision-making framework for the management of seriously ill newborns, developed by a Presidential Commission, provides physicians wide discretion based on their assessment of the benefits of treatment. In situations in which treatment is deemed futile, but the families request care, the Commission recommended providing treatment unless the provider declines to do so.[39]

In premature patients in whom further aggressive interventions were deemed to be futile, 67% of neonatologists surveyed reported adherence to these recommendations: foregoing care despite parental wishes for life-sustaining treatments.[40]

Numerous definitions have been offered for medical futility, and the legal system has given conflicting opinions regarding cases involving medical futility. Practitioners may consider an ethics consultation for cases of medical futility and should consult institutional policies and state laws.

The medical duty to sustain life is tempered by the ethical principle of nonmaleficence. It is therefore the physician's responsibility to advocate for withholding or withdrawing care when interventions are determined to be of no further benefit and may only bring harm to the patient.[41–44] It should be emphasized that the majority of pediatric intensivists in a survey do not favor unilateral "do not resuscitate orders" and instead prefer consultation with the family when the opportunity exists.[32] Some health care providers may have personal reasons that lead them to conscientiously object to withholding or withdrawing care. If the physician's objection to this course of action is a deeply held belief, he or she may also refer the patient to a provider who is willing to withhold or withdraw care as indicated.

Withdrawal or withholding of life-sustaining interventions for a child is a difficult and challenging decision for families. The current and future implications of this choice must first be thoroughly considered and discussed with the patient (as applicable) and family to determine their level of understanding, their wishes, their religious or cultural beliefs, and what to expect after withdrawal of life-sustaining treatment.[45,46] If the family and medical team decide to discontinue life-sustaining efforts, it is imperative to immediately initiate comfort care measures. These measures can include practices such as avoiding blood draws, invasive procedures, telemetry monitoring, and providing pain control as necessary.

Throughout this process, the needs of the family should be the primary focus. The family should be allowed to hold their dying child, if they wish, and be able to form a bond with the child by taking a photograph with them, or keeping a lock of hair or footprint sheet, and even naming the child. If the family wishes to have religious personnel, or extended friends and relatives, which they believe are essential for support during this difficult time, these people should be encouraged to visit as well. After the child has passed, supportive services should be offered to the family. Members of the medical team who provided treatment and established a rapport with the child and family may consider having a group meeting to discuss the events that took place and explain any questions or concerns they may have. These acts of compassion by the medical staff will forever be remembered by the family in the last moments of their child's life. Inclusion of patients' family members in making this decision is often beneficial to all parties involved.

In summary, after an initial resuscitation, multiple assessments of the patient should subsequently be performed to reevaluate prognosis and the prospective quality of life.[47] Therefore, further consideration of withdrawal of care must be made on a case-by-case basis.

CONCLUSIONS

It is vital for clinicians, and anesthesiologists in particular, to be aware of the ethical implications involved in the management of a neonate with unanticipated congenital malformations of the upper airway, which are likely lethal. Even with an unclear prognosis, initial resuscitation should be begun. Afterward, the physician can then reassess the patient's prognosis and have a discussion with family members to evaluate the benefits and harms of continued aggressive treatment.

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