

# Lethal

## Language,

# Lethal

## Decisions

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Although many of the congenital syndromes that used to be lethal no longer are, they are still routinely referred to as “lethal anomalies.” But the label is not only inaccurate, it is also dangerous: by portraying as a medical determination what is in fact a judgment about the child’s quality of life, it wrests from the parents a decision that only the parents can make.

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**N**eonatal and pediatric critical care have dramatically improved survival for children over the past twenty years. Neonatologists continue to reduce the gestational age and birth weight that a newborn must have achieved to survive, and advances in pediatric surgery and pediatric organ transplantation mean that complex congenital heart disease and biliary atresia no longer always result in death. They are no longer “lethal,” because with treatment, an infant can survive these conditions and even enjoy a good quality of life.<sup>1</sup>

Yet obstetricians, pediatricians, geneticists, and neonatologists continue to regard a category of congenital syndromes as “lethal anomalies.”<sup>2</sup> Examples are trisomy 13, trisomy 18, and anencephaly. Al-

though most professional discussions do not provide a specific definition of “lethal anomaly,” the term is generally used to refer to a child with (1) severe neurological compromise and (2) structural anomalies and/or functional disabilities that, if untreated, would cause death within a few months. The structural anomalies in these children include airway malformations, congenital heart disease, and gastrointestinal defects; the functional disabilities include swallowing dysfunction, aspiration, and apnea.

These anomalies and disabilities are usually treatable, and invariably are treated in an otherwise healthy infant. What makes them sometimes “lethal,” then, is the decision not to repair the anomalies or treat the disabilities in light of the child’s poor neurological prognosis. In effect, as we will argue, “lethal anomaly” is not an accurate clinical description; instead, it serves to convey an implicit normative view about quality of life.

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The hidden significance of the phrase is problematic because many parents may not rate the child's quality of life as dismally as their child's health care providers do. Data show that health care providers rank severely disabled health states lower than parents do,<sup>3</sup> which can partly explain why parents typically prefer more aggressive treatment than health care providers.<sup>4</sup> Data also show that what health care providers tell parents about a child's prognosis is influenced by the providers' attitude toward neurological impairment, with the effect that what is told to the parents depends on the provider.<sup>5</sup> Since this information influences family decision-making,<sup>6</sup> the phrase "lethal anomaly" obscures the normative nature of the decision and interferes with authentic parental autonomy.

### An Historical Overview

The practice of recommending that invasive treatments be withheld or withdrawn from infants with "lethal anomalies" first received critical attention in 1973, when Raymond Duff and AGM Campbell challenged the prevailing "social taboo" by making public and explicit their reasons for not providing invasive treatment to children with neurological disabilities.<sup>7</sup> They reviewed 299 consecutive deaths in the Yale Special Care Nursery and found that 43 (14 percent) were related to withholding treatment. They explained that the decisions were made by the physicians and parents because the child's prognosis "for meaningful life was extremely poor or hopeless"—a quality-of-life judgment that rarely had been publicly stated previously.<sup>8</sup>

Once the Pandora's box was opened, medical ethicists, theologians, and clinicians came out in support of and against this practice.<sup>9</sup> The dispute came to a head in the 1980s, when disability advocates and right-to-life groups publicized the issues surrounding the withholding of treatment, including nutrition, in handicapped newborns. They focused pub-

lic attention on the fact that hospitals and physicians were allowing children with congenital causes of developmental delay, such as Trisomy 21, to die from correctable birth defects. One such case to reach national attention was Baby Doe, who was born in 1982 with Trisomy 21 and a correctable gastrointestinal defect.<sup>10</sup> As a result of this and other cases, regulations were proposed that would have made withholding treatment for correctable birth defects illegal, regardless of the child's potential quality of life.<sup>11</sup> These regulations were successfully challenged on the ground that they were too restrictive.<sup>12</sup> Eventually, more modest regulations were passed as amendments to the 1984 Child Abuse Prevention and Treatment Act (often known as the Child Abuse Amendments or the CAA).<sup>13</sup>

Although the CAA did not dictate how physicians must practice, requiring instead only that the states develop a process by which to respond to cases of non-treatment, they were at least partly responsible for changing the standard of practice for treating mentally and physically handicapped newborns.<sup>14</sup> The CAA allowed for non-treatment in only a narrow range of cases: when the infant is chronically and irreversibly comatose; when the treatment would (1) merely prolong dying, (2) not effectively ameliorate or correct all of the infant's life-threatening conditions, or (3) otherwise be futile in terms of the survival of the infant; or if the provision of such treatment would be virtually futile or inhumane.<sup>15</sup> While the Federal Regulations explicitly stated that an anencephalic newborn would fit in these categories,<sup>16</sup> whether children with congenital anomalies like trisomy 18 and 13 met the criteria was unclear. Withholding treatment in children with trisomy 13 and 18 is still a common practice.

In the decades since Campbell and Duff, however, some families have begun to demand invasive treatment for these children despite medical recommendations for comfort measures only. Court cases have ensued when

physicians and parents have disagreed about the "futility" of treatment. A well-known example of such a case is Baby K of Virginia.<sup>17</sup> Baby K had anencephaly and was being cared for in a nursing home. On several occasions, she was transferred to the emergency room in respiratory failure, where her mother demanded mechanical ventilation. The physicians sought judicial relief from providing repeated mechanical ventilation that they deemed "futile." The court found for the baby based on the Emergency Medical Treatment and Active Labor Act (EMTALA), which does not allow a hospital to refuse life-saving treatment to anyone in extremis. Baby K lived for two and a half years, the longest that a child with anencephaly has ever survived. In fact, before Baby K, physicians had assumed an anencephalic child could not survive beyond one year. Clearly, the use of mechanical ventilation was not physiologically futile in this case; it successfully prolonged Baby K's life.

Part of the problem, then, is how to define futility. Mechanically ventilating a child with anencephaly is not physiologically futile because it will restore ventilation and prolongs the child's life. But it is futile if one uses a qualitative notion of futility; the child will never be able to interact with his or her environment, even if the child is weaned from the ventilator.<sup>18</sup>

Baby K is just one example of children with so-called lethal anomalies who have survived beyond infancy. Although these children will have severe neurological handicaps, they can and do achieve some minimal number of milestones.<sup>19</sup> If institutions aggressively treated all of these children who were born alive, the three- to five-year survival rate might even improve drastically, although survival into the teens would likely remain rare.<sup>20</sup>

## The Ethical Arguments

There are three main arguments against providing medical treatment for life threatening problems to *all* infants with “lethal anomalies.” The first is just that treatment is “futile,” but as we’ve noted, this claim depends on a conception of futility based on quality-of-life considerations and not on physiology. In many cases, treatment will not only address the clinical problem, but may result in prolonged life.

The second argument is that the burdens of treatment outweigh the benefits in these children. This claim is a value judgment and includes both the medical and psychosocial burdens and benefits. In general, parents are presumed to be the judge of whether the benefits of a treatment plan outweigh the burdens, all things considered. If the parents judge the treatments as more burdensome than beneficial, then they have the right to refuse further treatment. On the other hand, if the parents judge the treatments as beneficial, then their requests should be honored unless there is clear evidence that the judgment is mistaken. If the child appears to be experiencing significant unremitting pain or suffering, for example, then it is wrong to continue treatments that only prolong his survival and suffering. But rarely is this the case.

The third argument against treatment for life-threatening problems focuses on resource allocation and whether it makes economic sense to treat children with such a grim neurological prognosis when the money could be used for greater benefit elsewhere. The economic argument fails because the congenital anomalies classified as lethal are rare, so even if these children were given maximal treatment, their care would account for only a small percentage of the total U.S. health care budget. For example, Ramesh Sachdeva and colleagues found that only 1.6 percent of patient days in the PICU were spent on futile treatment of children

## It is not possible to ignore quality-of-life

who had either a “lethal anomaly” or a very poor quality of life secondary to PVS.<sup>21</sup>

But rejecting the resource question because it is not a true economic threat ignores the ethical issue in the resource allocation debate. The question is whether a quality-of-life criterion can be used as a threshold to determine whether treatment can be morally withheld. Although Duff and Campbell thought yes, the public outcry leading to the passage of the Baby Doe regulations showed that some disagreed vehemently.<sup>22</sup> In the absence of a social consensus to limit care, the practice has evolved to empower families, and not providers at the bedside, to make treatment decisions based on quality of life.

If these three arguments to withhold life-saving medical treatment from all infants with “lethal anomalies” fail, then what arguments would lead to providing the treatment? One is that physicians treat children with disabilities unjustly if they do not correct all life-threatening problems. This stance calls on physicians to treat all correctable medical conditions regardless of quality-of-life considerations and parental preferences—even to take legal custody if parents do not consent. This argument makes two assumptions: that it is possible to make medical decisions without incorporating quality-of-life judgments, and that it is appropriate to ignore quality-of-life judgments when a child’s neurological prognosis is dismal.

We reject these assumptions. First, it is not possible to ignore quality-of-life judgments when making medical decisions to withhold treatment without reverting to a vitalistic approach to medicine that we believe is untenable with modern technology.<sup>23</sup> Nancy Rhoden has shown that even the CAA exemptions incorporate quality-of-life judgments, although

**judgments when making medical decisions to withhold treatment without reverting to a vitalistic approach to medicine that we believe is untenable. Yet quality-of-life judgments are not only necessary; they are appropriate.**

narrowly circumscribed as exemplified by the permissibility of nontreatment of comatose infants who, with invasive treatment, might be able to live for years or decades.<sup>24</sup> Second, quality-of-life judgments are not only necessary; they are appropriate. As Rhoden has explained, consciousness is not valued as an end in itself, but as a condition for meaningful relationships. Meaningful relationships do not require “normal” or “near-normal” intelligence but only some ability to interact with others and with one’s surroundings. Without these abilities, many families decide their loved one would not want to continue to live.<sup>25</sup>

A second argument in favor of treatment is the belief that any life is better than death. Yet this argument also fails. While some health care providers, ethicists, and families believe that all life is sacred and should never be shortened,<sup>26</sup> their belief is but one of several possible ways of ordering the benefits and burdens; it should not be imposed on those who hold divergent beliefs.

If the arguments to provide treatment for all children with “lethal anomalies” fail, like the three arguments to withhold treatment, then

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the substantive question has to be regarded as an open question. The appropriate policy issue, then, may not be substantive but procedural: *who should decide* what is appropriate medical care for these children?

Throughout history, parents have been the primary decisionmakers for their children. Third-party intervention is restricted to cases in which parents are either abusive or neglectful of their responsibility. In caring for children with severe neurological compromise, there is no clear social consensus on whether treatment should be required or prohibited. Given this ambiguity, parental decisions to withhold or request treatment based on benefit-burden calculations should be respected.

Two caveats: First, some physicians, depending on their own religious or philosophical perspective, may find it difficult to comply with a decision to withhold treatment, and others may be uneasy about providing it. Yet respect for parental decisionmaking in such socially ambiguous circumstances should persuade physicians to put aside their own views when caring for such families. Ideally, then, the physician would ac-

cede to parental authority, provided that the decision is legally permissible. Alternatively, clinical responsibility could be transferred if the physician cannot come to terms with the family's decision.

Second, our arguments apply to treatment decisions for children with so-called lethal anomalies. What limits to family autonomy are appropriate when the neurological prognosis is less severe, or if a grim neurological prognosis is not associated with any other life-threatening conditions, are questions that go beyond the scope of this paper.

#### **Hidden Meaning**

Consider the case of Baby B, an eighteen-month-old infant with trisomy 18 admitted to the University of Chicago Pediatric Intensive Care Unit. Baby B had severe gastroesophageal reflux resulting in multiple episodes of aspiration pneumonia. He had not been gaining weight well, and his development was at the level of a two-month-old child.

Because of the child's "lethal anomaly," the child's primary physician believed that aggressive treatment of the pneumonias was inappropriate and wanted to place a do not resuscitate (DNR) order in the chart. The surgeons refused to perform a Nissen fundoplication and gastrostomy tube (which would reduce gastroesophageal reflux and so help avert aspiration pneumonia) because they believed it was "futile" to do so. But the mother wanted all medical treatment provided, including the surgery and mechanical ventilation, and refused to authorize a DNR order.

The normative nature of the decision not to treat a child like Baby B is evidenced by the tendency to talk about the treatment of pneumonia as "aggressive"<sup>27</sup> or to describe a fundoplication as "futile."<sup>28</sup> These procedures would be considered *routine* in an otherwise healthy infant. A gastrostomy tube and Nissen fundoplication is "medically indicated"<sup>29</sup> in a neurologically normal child with severe reflux; it becomes "aggressive" when the child has trisomy 18. Likewise, mechanical ventilation is "appropriate" for respiratory failure in an otherwise healthy child; it was "futile" for both Baby B and Baby K. The problem with such terms, then, is that they hide the quality-of-life judgments imbedded in the decision to withhold treatment, and thereby interfere with the parents' authority to define what is best for their child.

It has been almost three decades since Duff and Campbell made public that quality-of-life factors are integral to the decision to withhold or withdraw treatment in neonatal intensive care units. During this time, neonatal and pediatric intensive care practices have changed dramatically, both in their technical capabilities<sup>30</sup> and in their acceptance of greater parental participation in decision-making.<sup>31</sup> It is now time for the language to catch up. "Lethal anomalies" are no longer necessarily lethal, and should not be described as such. Frank discussions about quality of life are important, even if challenging, and can facilitate parental decision-making.

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#### **References**

1. J.D. Horbar et al., "Trends in Mortality and Morbidity for Very Low Birth Weight Infants, 1991-1999," *Pediatrics* 110 (2002): 143-51; W.T. Mahle, "Neurologic

and Cognitive Outcomes in Children with Congenital Heart Disease," *Current Opinion in Pediatrics* 13 (2001): 482-86; D.B. McElhinney and G. Wernovsky, "Outcomes of Neonates with Congenital Heart Disease," *Current Opinion in Pediatrics* 13 (2001): 104-110; and F.C. Ryckman et al., "Biliary Atresia-Surgical Management and Treatment Options as They Relate to Outcome," *Liver Transplantation and Surgery* 4, suppl. 1 (1998): S24-S33.

2. R.E. Behrman et al., eds., *Nelson's Textbook of Pediatrics*. 14th ed. (Philadelphia: WB Saunders, 1992), 284-86; K.L. Jones, ed., *Smith's Recognizable Patterns of Human Malformation*, 5th ed. (Philadelphia: W.B. Saunders, 1997), 20-21, 659-61; S.J. Hassed et al., "Perinatal Lethal Conditions: The Effect of Diagnosis on Decision Making," *Obstetrics and Gynecology* 82 (1993): 37-42; A.P. Bos et al., "Avoidance of Emergency Surgery in Newborns with Trisomy 18," *Lancet* 339 (1992): 913-15; and D.C. Wertz and M.B. Knoppers, "Serious Genetic Disorders: Can or Should They Be Defined?" *American Journal of Medical Genetics* 108 (2002): 29-35.

3. S. Saigal et al., "Differences in Preferences for Neonatal Outcomes among Health Care Professionals, Parents, and Adolescents," *JAMA* 281 (1999): 1991-97.

4. M.W. Doron et al., "Delivery Room Resuscitation Decisions for Extremely Premature Infants," *Pediatrics* 102 (1998): 574-82.

5. A.G. Randolph et al., "Factors Explaining Variability among Caregivers in the Intent to Restrict Life-Support Interventions in a Pediatric Intensive Care Unit," *Critical Care Medicine* 25 (1997): 435-9; and A.G. Randolph et al., "Variability in Physician Opinion on Limiting Pediatric Life Support," *Pediatrics* 103, no. 4 (1999): e46.

6. Randolph, "Variability," e46.

7. R.S. Duff and A.G.M. Campbell, "Moral and Ethical Dilemmas in the Special Care Nursery," *NEJM* 289 (1973): 890-94.

8. *Ibid.*, p. 893.

9. The major viewpoints are discussed in R. Weir, "Pediatricians and Selective Nontreatment" and "Options Among Ethicists," *Selective Nontreatment of Handicapped Newborns* (New York: Oxford University Press, 1984), 50-59, 143-87.

10. *In re Infant Doe*, No. GU8204-004A (Ind. Ct. App. 12 Apr 1982), *writ of mandamus dismissed bus nom. State ex rel Infant Doe v Baker*, No. 482 S. Ct. 27 May 1982, *cert. Denied*, 104 S. Ct. 394 (1983).

11. DHHS Notice to Health Care Providers, "Discriminating Against Handicapped by Withholding Treatment or

Nourishment," *Federal Register* 47, no. 116 (June 16, 1982) 26027; DHHS Interim Final Rule, "Nondiscrimination on the Basis of Handicap Relating to Healthcare for Handicapped Infants," *Federal Register* 48, no. 45 (5 July 1983), 30846-52; and DHHS Final Rule, "Nondiscrimination on the Basis of Handicap: Procedures and Guidelines Relating to Health Care for Handicapped Infants," *Federal Register* 49, no. 8 (12 January 1984), 1622-54.

12. American Academy of Pediatrics, National Association of Children's Hospitals and Related Institutions, *Children's Hospital National Medical Center, Plaintiffs v. Margaret M. Heckler*, 561 F. Supp. 395; 1983 U.S. Dist. LEXIS 17713 (1983).

13. Child Abuse Amendments of 1984 42 USCA 5101-5103 (1984).

14. See, for example, L.M. Kopelman, T.G. Irons, and A.E. Kopelman, "Neonatologists Judge the 'Baby Doe' Regulations," *NEJM* 318 (1988): 677-83; R.L. Ariagno, "Government Regulations in the United States" in *Ethics and Perinatology*, eds. A. Goldworth et al. (New York: Oxford University Press, 1995), 279-95; and I.D. Todres et al., "Life-Saving Therapy for Newborns: A Questionnaire Survey in the State of Massachusetts," *Pediatrics* 81 (1988): 643-49.

The change in neonatal practice can also be explained in part by the disability rights activists who successfully argued that nontreatment was discriminatory and by research that showed that many families with disabled children rated their lives quite positively. See P.M. Ferguson, A. Gartner, and D. K. Lipsky, "The Experience of Disability in Families: A Synthesis of Research and Parent Narratives" in *Prenatal Testing and Disability Rights*, eds. E. Parens and A. Asch (Washington DC: Georgetown University Press, 2000) 72-94.

15. See, child abuse amendments, 1984.

16. *Ibid.*, p. 5102.

17. *In the matter of Baby K*, 16F3d 590 (4th Cir. 1994). J.J. Paris, R. K. Crone, and F. Reardon, "Physicians' Refusal of Requested Treatment: The Case of Baby L," *NEJM* 322 (1990): 1012-1013.

18. S.J. Youngner, "Who defines futility?" *JAMA* 260 (1988): 2094-2095.

19. B.J. Baty, "Natural History of Trisomy 18 and Trisomy 13: II. Psychomotor Development," *American Journal of Medical Genetics* 49, no. 2 (1994): 189-94.

20. B.J. Baty, "Natural History of Trisomy 18 and Trisomy 13: I. Growth, Physical Assessment, Medical Histories, Survival, and Recurrence Risk," *American Journal of Medical Genetics* 49, no. 2 (1994): 175-88.

21. R.C. Sachdeva et al., "Resource Consumption and the Extent of Futile Care

among Patients in a Pediatric Intensive Care Unit Setting," *Pediatrics* 128 (1996): 742-7.

22. See Duff and Campbell, "Moral and Ethical Dilemmas," 1973.

23. N.K. Rhoden, "Treatment Dilemmas for Imperiled Newborns: Why Quality of Life Counts," *Southern California Law Review* 58 (1986) 1283-1347.

24. *Ibid.*, pp.1312-18.

25. *Cruzan v. Director Missouri Dept. of Health*, 58 U.S. L.W. 4916 (1990). *In re Quinlan*, 70 N.J. 10, 355 A.2d 647 (1976)

26. C.E. Koop, "The Sanctity of Life," *Journal of the Medical Society of New Jersey* 75 (1978): 62-9. See Weir, "Options among Ethicists."

27. J.C. Weeks et al, "Relationship between Cancer Patients' Predictions of Prognosis and Their Treatment Preferences," *JAMA* 279 (1998): 1709-14; and C. Hammerman et al., "Decisionmaking in the Critically Ill Neonate: Cultural Background v. Individual Life Experiences," *Journal of Medical Ethics* 23 (1997): 164-69.

28. R.S. Stein et al., "CPR-Not-Indicated and Futility," *Annals of Internal Medicine* 124 (1996): 75-7; E.M. Levine, "A New Predicament for Physicians: The Concept of Medical Futility, the Physician's Obligation to Render Inappropriate Treatment, and the Interplay of the Medical Standard of Care," *Journal of Law & Health* 9 (1994-5) 69-108; and L.J. Schneiderman, N.S. Jecker, and A.R. Jonsen, "Medical Futility: Its Meaning and Ethical Implications," *Annals of Internal Medicine* 112 (1990): 949-54.

29. The subjectivity of the concept "medically indicated" is discussed by Veatch in "The Concept of 'Medical Indications'" in R.M. Veatch *The Patient-Physician Relation: The Patient as Partner, Part 2* (Bloomington, Indiana University Press, 1991) 54-62.

30. See Horbar, "Trends in Mortality and Morbidity for Very Low Birth Weight Infants, 1991-1999," 2002. See also A.C. Chang, "Pediatric Cardiac Intensive Care: Current State of the Art and Beyond the Millennium," *Current Opinion in Pediatrics* 12, no. 3 (2000): 238-46; and J.M. Tilford et al., "Differences in Pediatric ICU Mortality Risk over Time," *Critical Care Medicine* 26 (1998): 1737-43.

31. H. Harrison, "The Principles for Family-Centered Neonatal Care," *Pediatrics* 92 (1993): 643-50; and N.E. Page and N.M. Boeing NM, "Visitation in the Intensive Care Unit: Controversy and Compromise," *AACN Clinical Issues in Critical Care Nursing* 5, no. 3 (1994): 289-95.