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THE AFTERMATH OF BABY DOE AND THE EVOLUTION OF NEWBORN INTENSIVE CARE

Mark R. Mercurio*

INTRODUCTION

It has been twenty-five years since the government regulations known as the “Baby Doe Rules” were put in place, largely in response to a widely publicized case in Bloomington, Indiana, in 1982.¹ In that case, a life-saving surgical procedure was withheld from an infant with Trisomy 21 (often referred to as Down Syndrome) and tracheoesophageal fistula, consistent with the parents’ wishes and the advice of at least one of their physicians. A court ruling upheld the parents’ right to refuse the surgery, and the infant subsequently died.²

This review is presented as one of several to be discussed and published together as part of a symposium in which these rules, and the events that have followed, are to be evaluated from legal, medical, and ethical perspectives. This particular essay is intended to provide an overview of the evolution of the field of newborn intensive care since the 1980s, particularly as it relates to practices surrounding withdrawal or withholding of life-saving or life-sustaining treatments. Before discussing selected clinical issues and dilemmas of the past quarter century, some relevant historical background leading up to the Baby Doe case is presented. The Baby Doe Rules themselves will not be discussed here in any depth, as that will be covered by the speakers (authors) that follow.

Presentation of clinical developments and ethical dilemmas will be followed by an overview of the evolution of specific ethical

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1. Child Abuse Amendments of 1984, Pub. L. No. 98-457, 98 Stat. 1749 (codified as amended at 42 U.S.C. §§ 5101–5106i (2006) and implemented in relevant part by 45 C.F.R. § 1340.15 (2008)).

2. *In re Infant Doe*, No. GU8204-004A (Monroe County Cir., Ind. Apr. 12, 1982) (deferring to parental decision to withhold treatment for a disabled newborn), *cert. denied*, 464 U.S. 961 (1983).

principles and questions that have been influential in the field of neonatology. Again, more in-depth discussions of these will be presented by the speakers that follow. This essay is intended to provide a backdrop for those discussions.

I. HISTORICAL BACKGROUND

The Baby Doe case, the rules that followed, and the evolution of thinking with regard to withholding certain treatments from newborns are all best considered in light of events that went before. Obviously, a thorough discussion of the relevant philosophical thought, medical practice, and legal precedent is well beyond the scope of this essay. Nevertheless, a few landmark events and ideas are worthy of at least brief mention. Thus, though this paper is intended to discuss the aftermath of the Baby Doe Rules, some time is first devoted to ideas and events that preceded them.

A. Patient Autonomy and the Legacy of Nuremberg

The evolution of medical ethics, and medical practice, has been influenced by many historical and political events, but few, if any, have had as much impact as the events in Europe during the Nazi era. Medicine in the years since the mid-twentieth century has been profoundly affected by what was carried out not only by politicians and soldiers but by members of the medical profession as well.

The concept of individual autonomy surely predates that era, as found in the words of John Stuart Mill in his nineteenth-century essay *On Liberty*: “Over himself, over his own body and mind, the individual is sovereign.”³ In the medical arena, in the early twentieth century, Justice Benjamin Cardozo applied this same concept to a patient’s right to refuse a surgical procedure: “Every human being of adult years and sound mind has a right to determine what shall be done with his own body; and a surgeon who performs an operation

3. JOHN STUART MILL, *ON LIBERTY AND OTHER ESSAYS* 14 (John Gray ed., Oxford Univ. Press 1998) (1991).

without his patient's consent commits an assault, for which he is liable in damages."⁴ Nevertheless, medical practice remained widely influenced by paternalistic approaches, and the relatively recent dominance of patient autonomy among our guiding principles is arguably a result, at least in large part, of the atrocities committed by some German physicians during the Nazi era.

1. The Code of Nuremberg and the Right to Refuse

In 1947, in the wake of the Second World War, and in response to revelations regarding atrocities carried out in name of medical research, the war crimes tribunal developed the Code of Nuremberg. This code was intended to protect human subjects of medical experimentation. Ten standards for physicians carrying out such research were presented, and the first began with the words: "The voluntary consent of the human subject is absolutely essential."⁵ This idea has subsequently been extended to clinical medicine and the doctrine of informed consent for medical treatments. Thus, it is widely accepted in the United States that adult patients of sound mind generally have a right to refuse medical treatment, even life-saving treatment. It has further been extended, in most circumstances, to parents, affording them to right to refuse medical treatment on behalf of their children. However, it is generally accepted within the pediatric profession that the parental right to refuse treatment, particularly life-saving treatment, is more limited than an adult's right to refuse on his own behalf, as will be discussed below.

2. Medical Science Under Dictatorship

Leo Alexander, M.D., served as a consultant to the U.S. Secretary of War, on duty with the Office of the Chief Counsel for War Crimes at Nuremberg. His discussion of the atrocities of that era, and the

4. *Schloendorff v. Soc'y of N.Y. Hosp.* 105 N.E. 92, 93 (N.Y. 1914).

5. Jochen Vollman & Rolf Winau, *Informed Consent in Human Experimentation Before the Nuremberg Code*, 313 BRIT. MED. J. 1445, 1448 (1996), available at <http://www.cirp.org/library/ethics/nuremberg/>.

complicity of physicians, were documented in a landmark article in *The New England Journal of Medicine*.⁶ Perhaps most relevant to this essay, and to any discussion regarding withholding treatment from some newborns, is his observation regarding the beginnings of those crimes: “It started with the acceptance of the attitude, basic in the euthanasia movement, that there is such a thing as a life not worthy to be lived.”⁷ This concept, or, perhaps variations of it, is central to the controversy of Baby Doe, and the regulations and clinical practices that followed.

B. Withholding Treatment from Newborns in the United States

The practice of withholding care from malformed infants, as well as infanticide, dates back at least to ancient Greece and Rome.⁸ Widespread discussion in the United States specifically regarding withholding life-saving procedures from infants with Trisomy 21 predates the *Bloomington* case, as shown below. Once again, an exhaustive discussion of the relevant cases and literature is beyond the scope here, but a few important events in the years leading up to 1982 deserve mention.

1. The Johns Hopkins Cases, 1971

In 1971, at Johns Hopkins Hospital in Baltimore, a baby presented with Trisomy 21 and duodenal atresia. Trisomy 21 is the presence of an extra chromosome 21 in every cell that results in a characteristic facial appearance (leading to the term “mongolism” used at that time), mental retardation, and some combination of several other health problems and birth defects. Duodenal atresia is a complete obstruction of the small intestine, present at birth, that prevents the passage of liquid or solid food. Children with duodenal atresia in 1971 routinely underwent surgical repair, with generally favorable results. In this case the parents refused to give consent, the surgeon

6. Leo Alexander, *Medical Science Under Dictatorship*, 241 NEW ENG. J. MED. 39, 44 (1949).

7. Alexander, *supra* note 6.

8. GREGORY PENCE, CLASSIC CASES IN MEDICAL ETHICS 216 (McGraw-Hill, 4th ed. 2004) (1990).

concluded, the case did not go to court, and the child died. A second child with Trisomy 21 and a similar obstruction at Johns Hopkins that year was also allowed to die after parents refused surgery. The mother cited concerns about the burden the infant would impose on the other members of the family.⁹ These cases raised considerable attention, at least within the pediatric profession.

2. *Duff and Campbell's Landmark Article, 1973*

In 1973, Drs. Raymond Duff and Alex Campbell at Yale-New Haven Hospital published a landmark article in *The New England Journal of Medicine*, in which they documented the fact that, in their hospital, babies were often allowed to die. They reviewed 299 consecutive infant deaths from 1970 to 1972, and reported that 43 (14%) were related to withholding treatment. They reported, and strongly endorsed, joint decisions made by physicians and families to withhold treatment. In addition, they noted that some families specifically cited their own burden in raising a handicapped child as a rationale for the decision to withhold treatment.¹⁰ Duff and Campbell clearly felt that parents had a right to make such decisions based on the interests not just of the infant patient but also considering their own interests and those of their other children. Their paper gained a great deal of attention, and stirred debate, in large part because, in their words, they had broken "professional silence . . . on a major [] taboo"¹¹

3. *David Smith and the Infant's Best Interest, 1974*

A different point of view regarding such cases was published the following year in the medical ethics literature by a (then) little-known junior faculty ethicist at, co-incidentally and interestingly, Indiana University in Bloomington. Almost certainly it was read by a small

9. *Id.* at 217–18.

10. Raymond S. Duff & A.G.M. Campbell, *Moral and Ethical Dilemmas in the Special-Care Nursery*, 289 *NEW ENG. J. MED.* 890, 890–91, 894 (1973).

11. *Id.* at 894.

fraction of those who read Duff and Campbell's article the year before, and its immediate influence on medical practice is difficult to discern. It is nevertheless worthy of note for at least three reasons: (1) David Smith subsequently became a major figure in the field of bioethics, (2) it is a fascinating coincidence of history that the Baby Doe case would play out eight years later in the same town where Smith lived (the principal physicians in the 1982 case were both acquaintances of his), and (3) his essay eloquently argues for a different approach than that taken in the landmark cases that occurred both before and after it was written. Smith argued that "the only fair criterion for deciding appropriate treatment for a given baby is that baby's own welfare and ability to receive love."¹²

Smith's argument was influenced by those of by his mentor, Paul Ramsey, at Princeton. It stands in clear distinction to Duff and Campbell, who argued that interests of other individuals, in addition to those of the infant patient, could and should be considered when deciding whom to treat. This remains, to this day, a central question in medical ethics, and specifically in the ethical discussions surrounding withholding treatment from certain infants. Should such decisions be made solely based on the interests to the infant patient, assessed by weighing the benefits and burdens of the proposed treatment and ongoing life? Or, can decision makers also consider the interests of others, such as family members?

4. *Physician Attitudes in the 1970s*

There is considerable evidence of support among physicians in the 1970s for decisions similar to those made at Johns Hopkins. A survey reflecting the views of 230 Massachusetts pediatricians, for a hypothetical case of Trisomy 21, duodenal atresia, and parental refusal of surgery, was published in 1977. Surgery was recommended by 46% of pediatricians surveyed. That is, most would not recommend it in the face of parental refusal. Of those who did

12. David H. Smith, *On Letting Some Babies Die*, 2 HASTINGS CENTER STUD. 37, 45 (1974).

recommend surgery, only 40% would pursue a court order. When asked if the presence of mental retardation or severe physical malformation justified withholding consent for life-saving procedures for psychosocial reasons, such as disrupting the marriage or effect on other siblings, an equal number felt that psychosocial reasons should and should not be considered.¹³ A larger national survey (457 physicians), also published in 1977, posed essentially the same question. Here it was found that 50% of pediatricians, geneticists, and neonatologists would acquiesce to a parental refusal of surgery, and 77% of surgeons would acquiesce.¹⁴

It is in this light that one must consider decisions made by physicians and parents in 1973, and in the subsequent Bloomington Baby Doe case of 1982. At the very least, it can be stated that decisions to withhold treatment in these highly publicized cases did not represent clear deviation from standard medical care at the time.

5. *The Case of Baby Doe, Bloomington, 1982*

This takes us to the well-known case that became the impetus for the regulations that followed. In 1982 at Bloomington Hospital in Bloomington, Indiana, an infant was born with Trisomy 21 and tracheo-esophageal atresia (TEF). TEF is a birth defect that usually includes an obstruction of the esophagus, as well as an abnormal connection between the esophagus and trachea. As with duodenal atresia, the infant is unable to take in liquid or solid food without surgical repair. Thus the situation was very similar to the previous cases discussed. The infant's physician recommended transfer to Riley Children's Hospital in Indianapolis for surgical repair. The obstetrician, on the other hand, felt that non-intervention would be preferable, and the parents agreed and refused surgery. Hospital administrators and pediatricians disagreed, and an emergency session

13. I. David Todres et al., *Pediatricians' Attitudes Affecting Decision-Making in Defective Newborns*, 60 PEDIATRICS 197, 200 (1977).

14. Anthony Shaw et al., *Ethical Issues in Pediatric Surgery: A National Survey of Pediatricians and Pediatric Surgery*, 60 PEDIATRICS 588, 590 (1977).

was convened with the county judge. The court upheld the parents' right to refuse the surgery, and this ruling was upheld on two subsequent appeals. The infant died before the case could be heard in the U.S. Supreme Court.

Among the most notable and troubling aspects of this case from a physician's point of view is the testimony of the obstetrician that even if the surgery were successful "the possibility of a minimally adequate quality of life was non-existent [due to] the child's severe and irreversible mental retardation."¹⁵ This is inconsistent with what is known now, and was known then, about the degree of cognitive disability associated with Trisomy 21.

6. *Baby Doe Regulations*

The Bloomington case generated a great deal of public attention, and led to swift action by the federal government in an attempt to prevent similar cases. The result was a series of regulations and court cases that are better discussed by those with expertise in the law, and will be the focus of much of the discussion in the other sessions of this symposium. Briefly, it is noted that only one month after infant Doe's death the first of these regulations was issued by the federal government, prohibiting discrimination based solely on handicap. Posters were placed in NICUs (neonatal intensive care units) warning against discriminatory failure to feed and care for handicapped infants, a toll-free number was provided to report such violations, and "Baby Doe Squads" were established to respond to such reports. In the nineteen months that the squads were active (1982–1983), they received 1,633 calls, investigated forty-nine, and appear to have had an effect on treatment in six.¹⁶ It remains unclear whether the regulations currently in place are consistent with common American pediatric practice or the generally accepted medical standard that decisions about withholding treatment in infants should be determined by an assessment of the infant's best interests. This

15. PENCE, *Supra* note 8, at 220.

16. *Id.* at 221–22.

potential inconsistency was raised in 2005 by Professor Loretta Kopelman¹⁷ and was discussed by her later in this symposium.

7. *President's Commission*

The President's Commission for the Study of Ethical Problems in Medicine and Biomedical and Behavioral Research released a report in 1983 regarding ethical issues surrounding decisions to forego life-sustaining treatment. The Commission concluded that in some cases withholding such treatment was appropriate. The Commission was also critical of the approach taken by the Baby Doe Regulations then in effect and instead recommended that hospitals should develop mechanisms, such as ethics committees, to oversee and review decisions to withhold life-sustaining treatments.¹⁸

8. *Withholding Treatment in the Years Since Baby Doe*

In the years since the Baby Doe case and the subsequent federal regulations, physicians have continued to withhold treatments, including life-sustaining medical treatments, from some newborns. One review of 165 deaths in the NICU at the University of California at San Francisco Medical Center from 1989 to 1992 showed that 108 deaths were the result of withdrawal of life-sustaining therapy. An additional thirteen deaths were due to withholding additional therapy.¹⁹ This author can speak from personal observation and experience, as well as numerous conversations with neonatologists from throughout the United States, that this practice was and is widespread. It remains widely believed among neonatologists that in certain cases parents have the right to refuse medical interventions including surgery, resuscitation, mechanical ventilation, and other

17. Loretta Kopelman, *Are the 21-Year-Old Baby Doe Rules Misunderstood or Mistaken?*, 115 PEDIATRICS 797, 797-98 (2005).

18. PRESIDENT'S COMMISSION FOR THE STUDY OF ETHICAL PROBLEMS IN MEDICAL AND BIOMEDICAL AND BEHAVIORAL RESEARCH, *DECIDING TO FOREGO LIFE-SUSTAINING TREATMENT* 219, 224, 227 (1983).

19. Stephen N. Wall & John C. Partridge, *Death in the Intensive Care Nursery: Physician Practice of Withdrawing and Withholding Life Support*, 99 PEDIATRICS 64, 64, 66 (1997).

life-sustaining medical treatments. Furthermore, as will be shown in the final section of this essay, that belief is consistent with guidelines published by the American Academy of Pediatrics.

That is not to say, however, that things have not changed. Notably, while neonatologists generally support the parental right to refuse treatment in certain situations, the threshold for that right appears to have moved. Specifically, in the case of Trisomy 21, the standard of care for many years has now been to provide surgical correction of intestinal atresias or heart disease, and parental refusal of such treatment would most likely be challenged in court and overruled. This may reflect a diminution of perceived parental authority or rights, but it almost certainly reflects a clearer understanding of the prognosis for “quality of life” for people with Trisomy 21. The Baby Doe case could well be seen as good ethical reasoning (the parents’ right to refuse treatment when the prognosis is extremely bleak) applied to bad data (the quality of life in Trisomy 21 was far less bleak than stated). One challenge for the present and future, then, is to be certain that parents and physicians make such decisions with the best possible understanding of available outcome data for the situation at hand.

II. THE EVOLUTION OF CLINICAL PRACTICE

The evolution of newborn intensive care over the past quarter century has been marked by many changes. Most visible perhaps are the physical changes. Neonatology is now practiced in far more settings, and by far more individuals. In the state of Connecticut, for example, the number of NICUs has doubled. This trend has been seen on a national level as well. The intensive care units are larger, more spacious, and the monitors are also larger, with striking color displays. There are many more of the smallest babies, as will be discussed below. Moreover, there are many more neonatologists. In 1985 neonatology was a relatively new subspecialty within pediatrics. As NICUs in the major academic centers continued to train neonatology fellows over the past twenty-five years, the number

of trained neonatologists has grown accordingly, and this may in part have fueled the growing number of NICUs found throughout the country.

A. Technologic Advances

There have of course been many technological advances, but it is worth reviewing some whose progress is particularly interesting in light of the ethical questions raised by the case of Baby Doe and others like it.

1. Prenatal Diagnosis

The practice of neonatology has been significantly affected by the dramatic increase in the effectiveness of, and use of, techniques for prenatal diagnosis. Perhaps most important among these is prenatal ultrasound. In the past quarter century we have seen remarkable improvements in the resolution of this test, as well as its near universal use in pregnancy in the United States. Congenital anomalies that in the past usually presented at birth, or in the first days of life, are now commonly diagnosed prenatally. This includes diagnoses such as congenital heart disease, intestinal atresias, and tracheoesophageal atresia, among many others. Even Trisomy 21 is routinely diagnosed prior to birth, due in part to the ability to pick up abnormalities on ultrasound as subtle as thicker skin around the neck, as well as chromosomal screening by amniocentesis.

The result has been fewer surprises for the obstetricians, parents, and neonatologists—though they still occur. When a significant prenatal diagnosis is made, mothers usually have the option to deliver at a center optimally equipped to deal with the anticipated problem. Were this technology as sophisticated and widespread in 1982, Baby Doe's Trisomy 21, TEF, or both would likely have been diagnosed or at least suspected before birth. It is interesting to speculate how that information might have affected the course of events.

In addition to fewer surprises, and a far better ability to prepare for a child with significant anomalies, there is another very important

change in newborn intensive care that has resulted from improved prenatal diagnosis: a notable decrease in some very bleak disorders, due to termination of pregnancy. For example, anencephaly (a lethal congenital defect that involves absence of most of the brain) is seen far less frequently in the NICU, as these are routinely diagnosed on prenatal ultrasound and most often the pregnancy is terminated.

All of this is quite relevant to the ethical discussions at hand, because these patients who receive a worrisome or ominous antenatal diagnosis are often the ones for whom neonatologists and parents struggle to find the best course of action. With a diagnosis before birth, the discussion is thus moved back, and the obstetrician often plays a central role. Furthermore, depending on the timing of diagnosis, termination of pregnancy is often an option available to the pregnant woman, whereas euthanasia after delivery is not. Here again is seen the importance of every physician involved in such cases being knowledgeable about the outcome for the diagnoses at hand and presenting the information in a clear and thorough manner. Obstetricians commonly (and rightly) involve neonatologists, geneticists, and pediatric surgeons in counseling women in these situations.

2. Assisted Reproductive Technology

Assisted reproductive techniques, such as in vitro fertilization, have greatly advanced the available options and the likelihood of success for previously infertile couples. How has this changed the field of neonatology? One aspect of in vitro fertilization is the ability to implant several embryos at one time into a woman, in the hopes that one or two will be successful. There are, to my knowledge, no legal restrictions in the United States on this practice. Sometimes the result is a successful singleton birth, but often the result is multiple gestation. Years ago triplets were an extraordinarily rare phenomenon, but far less so today as a result of this technology, and of course quadruplets and beyond have occurred as well. At the time of this writing, it has just been announced in the media that a woman in California has successfully delivered octuplets.

Multiple gestations carry with them an increased risk of prematurity, and all of the problems that come with it. This technology is relevant to the discussion for at least three reasons. First, it has fueled the national debate regarding the moral status of embryos, as some embryos created in this process could be discarded, donated, or used for research. Second, it has raised much discussion within the profession about the ethics of producing multiple gestations, sometimes referred to in the media as “miracle babies,” who may be at increased risk for permanent neurological and other sequelae as a result of extreme prematurity. The more fetuses present in a uterus, the earlier delivery is likely to occur, though wide variation is seen. Finally, it has increased the number of preterm babies that occupy NICUs, and, as will be discussed next, preterm birth, particularly at borderline viability, has become a central point of ethical discourse within neonatology.

3. *Extreme Prematurity*

Over the past quarter century the survival of the tiniest babies has improved significantly. Tyson et al. and The National Institute of Child Health and Human Development (NICHD) recently published survival and morbidity data for more than 4,000 babies born at nineteen major academic medical centers in the United States from 1998 to 2003.²⁰ From this publication and others, it is evident that some newborns commonly believed non-viable in 1985, such as those below 500 grams or below twenty-three weeks gestation, are now surviving. Some of this improvement is likely due to improved treatments for respiratory disease in premature infants, such as surfactant replacement therapy, and better mechanical ventilation of the smallest newborns. The lower limit of viability has moved somewhat, but most babies at or near the limit will not survive despite our best efforts, and many of the survivors will have severe permanent neurologic sequelae such as cognitive disability, cerebral

20. Jon E. Tyson et al., *Intensive Care for Extreme Prematurity—Moving Beyond Gestational Age*, 358 NEW ENG. J. MED. 1672, 1673–75 (2008).

palsy, blindness, and hearing deficits. The basic ethical problem of prematurity that plagued parents and neonatologists in 1985 remains: At what gestational age or size does it become acceptable to withhold resuscitation and intensive care?

The American Academy of Pediatrics (AAP) Neonatal Resuscitation Textbook provides the following guidance to neonatologists:

Where gestation, birth weight, and/or congenital anomalies are associated with almost certain early death, and unacceptably high morbidity is likely among the rare survivors, resuscitation is not indicated, although exceptions may be appropriate in specific cases to comply with parental request. Examples may include . . . confirmed gestational age of less than 23 weeks or birth weight less than 400 gm.²¹

This advice leaves much room for subjective judgment as to what is “almost certain” or “unacceptably high,” but nevertheless provides some framework upon which to craft a reasonable decision.²²

The Tyson paper and the NICHD data, however, suggest that it makes little sense to make a decision regarding resuscitation based on gestational age alone. Within a given gestational age the chances of survival and disability vary widely, influenced by other factors including size, gender (preterm girls do better than boys), antenatal treatment with steroids, and single versus multiple gestation (singletons do better than twins or triplets, for instance). Moreover, the NICHD has provided an online tool²³ that enables physicians to enter these variables and learn the likelihood of survival and disability. The tool is of course not perfect, but represents a major step forward in approaching this difficult question. It enables us to give parents a more accurate assessment of their child’s chances.

21. TEXTBOOK OF NEONATAL RESUSCITATION 9–5 (American Academy of Pediatrics, 5th ed. 2006) [*hereinafter* AAP TEXTBOOK].

22. *Id.*

23. http://nichd.nih.gov/about/org/cdbpm/pp/prog_epbo_case.cfm.

Some ethical blunders in the past may well have been based on a poor understanding of the prognostic data. Accurate data alone do not provide the best ethical answer, but they are an essential ingredient.

As a result of the NICHD data, the conversation may eventually move from, “Below what gestational age is it acceptable not to resuscitate?” to, “Below what chance of survival, or survival without severe disability, is it acceptable to resuscitate?” The fundamental ethical questions, however, remain largely unchanged: Should parents and physicians consider the interests of the family, or should the decision be based solely on the interests of the child? How much choice should parents be given? At what point should physicians seek to overrule a parental decision? Should these questions be viewed differently for infants than for older children or adults? If so, what is the justification?

4. Hypoplastic Left Heart Syndrome

Hypoplastic left heart syndrome (HLHS) is a term used to describe underdevelopment of the left side of the heart, uniformly fatal if left untreated. The nature of fetal circulation is such that a fetus can grow and develop well with this abnormality, relying primarily on the right side of the heart, and appear normal at birth. He or she may appear normal for one or several days, relying on the fetal circulation. Infants with normal hearts make the transition from fetal to neonatal circulation over the first one to two days (where the left side of the heart takes over an essential role) without difficulty. The infant with HLHS, however, will go into cardiovascular shock when the fetal circulation closes and the underdevelopment of the left side becomes evident.

In the 1980s these patients often presented, tragically, as “normal” full term babies in the well-newborn nursery who at one to two days of age became quickly and tragically ill. Some presented to the emergency department shortly after discharge home. Surgical interventions were being developed, but the overall chance of success was poor. Transplant was an option, though infant donor hearts were infrequently available. So desperate was the situation that one patient

at Loma Linda University was transplanted with a baboon heart in 1984 and died after twenty days. When counseled about the poor outlook, parents commonly asked for comfort measures only, and physicians readily complied. The chance of successful treatment was felt to be so poor that there was little dissent within the profession about this approach.

Important changes have occurred since the 1980s with regard to HLHS. First, these infants are now commonly diagnosed on prenatal ultrasound. As a result, fewer present unexpected as newborns. Women may elect to terminate the pregnancy, and if they choose to deliver and receive aggressive surgical management, delivery is arranged at a center where such treatment is available. Second, the efficacy of surgical repair has risen dramatically during the past two decades. Standard surgical treatment involves three separate operations over (at least) three separate hospitalizations. This surely represents a great deal of hardship for the child and the family, but now more than one-half of these children can survive for at least several years and likely well beyond that.

The likelihood of survival with aggressive surgical treatment has led many physicians involved (neonatologists, pediatric cardiologists, and pediatric cardiovascular surgeons) to view treatment as obligatory rather than optional for parents. The burden of treatment, the still considerable mortality, and the long-term neurological problems seen in many of the survivors have led some other physicians to feel that non-treatment (comfort care only) should remain an option available to the parents. This stands at the time of this writing as a significant ethical disagreement among qualified, thoughtful physicians on both sides of the issue. At its core, it is the same question faced in the 1980s with regard to Baby Doe and others, and faced both then and today with regard to extreme prematurity: How low must the odds of survival be, or how severe

must the burdens be, to justify allowing parents to choose non-treatment?²⁴

B. Changing Approaches to Aneuploidy

Aneuploidy refers to the condition of having an abnormal number of chromosomes, which invariably results in physical abnormalities to some degree. Trisomy 21, also known as Down Syndrome, is the most common aneuploidy among live births. As discussed above, it can be associated with a variety of anomalies such as intestinal obstruction, structural heart defects, and many others, but invariably includes facial changes and some degree of cognitive disability. The next most common aneuploidy among live births is Trisomy 18, then Trisomy 13. Each of these can also result in a variety of anomalies, but each invariably includes profound mental deficits, far worse than that seen in Trisomy 21, and very shortened life spans.

1. Trisomy 21

A notable example of the changing views within neonatology with regard to Trisomy 21 can be found in the interesting case of extracorporeal membrane oxygenation (ECMO), a technology used to treat some infants with extremely severe but reversible lung disease. ECMO is essentially a form of bypassing the lungs, using an external machine to oxygenate the blood. It is commonly carried out for seven to ten days, is extremely expensive and labor-intensive, and is among the most intensive measures offered in any NICU. For example, at the time of this writing there are only two NICUs in New England capable of providing ECMO.

Some patients are not considered candidates for this procedure even if their lung disease warrants it. They may be too small for the equipment available, or have another disorder that makes it unlikely ECMO would be successful. In the 1980s and 1990s, Trisomy 21 was

24. See generally Mark R. Mercurio et al., *Left Heart Syndrome, Extreme Prematurity, Comfort Care Only, and the Principle of Justice*, 122 PEDIATRICS 186 (2008).

generally considered a contra-indication for ECMO. Therefore, parents of infants with Trisomy 21 whose severity of respiratory disease otherwise warranted ECMO were generally not offered it, despite the fact that there was no evidence that these children were less likely to respond to the therapy. This thinking appears to have evolved. In a recent survey of ECMO centers nationally and internationally, 69% of respondents say they usually or always offer ECMO to patients with Trisomy 21 who otherwise meet criteria, though 12% responded rarely or never.²⁵

This specific example reflects a more general trend with regard to the approach to patients with Trisomy 21. It is now considered the standard of care to provide these children with the same degree of aggressive medical and surgical intervention as would be offered those without any known chromosomal abnormalities. In our NICU today, if a parent of a child with Trisomy 21 were to refuse repair of duodenal atresia or tracheoesophageal fistula, a court order would be sought and almost certainly obtained.

2. *Trisomy 18 and 13*

Most patients with Trisomy 13 or 18 die in the first month of life, and fewer than 10% survive the first year. Their extremely poor prognosis, as well as their profound cognitive deficits, have generally led neonatologists to limit available treatments, such as surgical correction of structural heart disease commonly associated with these trisomies. While all survivors will be severely affected mentally, it is not really clear what percent would survive if all of these infants received aggressive medical and surgical care. There is perhaps something of a self-fulfilling prophecy at work when one looks at survival data for such infants. Most die in the first month; therefore we do not try aggressively to keep them alive, and as a result, most die in the first month. This begs three important questions: (1) What

25. Rachel L. Chapman, Steven M. Peterec, Mathew J. Bizzarro, & Mark R. Mercurio, *Patient Selection for Neonatal Extracorporeal Membrane Oxygenation: Beyond Severity of Illness*, 29 J. PERINATOLOGY 606, 606–11 (2009).

would be the predicted survival and lifespan if we tried to save these children? (2) Should we offer parents aggressive care? (3) Are we making the same ethical mistake that was made in the past regarding Trisomy 21?

Congenital heart disease occurs in 60-90% of these patients. Though in most cases over the past two decades (and before) cardiac surgery was not offered to these families, it has occurred. In 2004 the Pediatric Cardiac Care Consortium reported data on infants and children with cardiac anomalies since 1982, including more than 83,000 cases in North America and Europe. Of these, they identified thirty-five patients with Trisomy 13 or 18 who underwent cardiac surgery. Survival to hospital discharge was 91%, suggesting that these patients can tolerate the surgery, but no long-term survival data were provided.²⁶ More recently, Japanese authors have reported that aggressive cardiac management (including surgical repair) to patients with Trisomy 13 and 18 results in a longer survival times and advocate for offering this approach to families. Because some of their patients were alive at the time of data acquisition, the exact effect on lifespan is not clear.²⁷

What is clear, however, is that some physicians are now offering cardiac surgery to these families. Cardiac surgery on a patient with Trisomy 13 and ventricular septal defect (VSD) was carried out for the first time at Yale-New Haven Children's Hospital in 2005, after a strong request by parents and review with the hospital ethics committee. The question of whether surgery should even be made available to parents remains a point of disagreement among physicians, and to my knowledge no physician at this hospital or elsewhere has ever considered it mandatory. That is, neonatologists and cardiologists in this institution (and to the best of my knowledge elsewhere in the United States) would respect parental refusal of surgery.

26. Eric M. Graham et al., *Effectiveness of Cardiac Surgery in Trisomies 13 and 18 (from the Pediatric Care Consortium)*, 93 AMERICAN J. CARDIOLOGY 801, 801-02 (2004).

27. Yukihiro Kaneko et al., *Intensive Cardiac Management in Patients with Trisomy 13 and Trisomy 18*, 146A AM. J. MED. GENETICS PART A 1372, 1372, 1379 (2008).

How does this differ from decisions in the past regarding Trisomy 21? Here again, we see that the line has moved, but the fundamental issue is the same. Neonatologists allow parents to decline surgery or intensive care measures, it is here argued, not solely based on the poor lifespan. Note that we do not really know what the lifespan would be with aggressive management, but the limited data available suggest it can be extended, and some survive for years. Rather, it is here suggested that aggressive treatment is considered optional (if offered at all) because of the profound neurological disability in these children and concerns about quality of life. There are also concerns about burdens to the family, which some ethicists and physicians will find relevant, and some will not.

The AAP textbook on resuscitation has given advice on this matter similar to that for patients below twenty-three weeks and 400 grams:

Where gestation, birth weight, and/or congenital anomalies are associated with almost certain early death, and unacceptably high morbidity is likely among the rare survivors, resuscitation is not indicated, although exceptions may be appropriate in specific cases to comply with parental request. Examples may include . . . confirmed gestational age of less than 23 weeks or birth weight less than 400 gm.²⁸

The problem is “almost certain early death” might be a bit less certain with aggressive care.²⁹ There are also, however, other AAP guidelines (reviewed below) that more clearly emphasize the importance of the patient’s best interest, and weighing the perceived benefits and burdens to the patient of the proposed treatment. It is here suggested that the vast majority of neonatologists (including this author) would say that such an analysis permits non-treatment. One might also wish to consider how non-treatment in this setting conforms to the words and spirit of the Baby Doe Regulations.

28. AAP TEXTBOOK, *supra* note 21.

29. *Id.*

Professor Kopelman might suggest that it does not, and that the problem is not with the practice but rather with the wording of the regulations, and with the AAP's tacit support of those regulations.

Also, even among those who advocate for "comfort care only" for infants with trisomy 13 or 18, it has been suggested that certain surgeries, such as the relatively simple and highly successful repair of VSD, should be considered a component of comfort care. That is, by relieving the respiratory distress that eventually accompanies a large VSD, the patient is made more comfortable even during his likely shortened lifespan. If this premise were accepted, one might argue that such surgery should be mandatory rather than optional for parents. The counter-argument might be that the respiratory discomfort brought on by the VSD could be treated with less invasive means, such as the use of morphine. In any case, the argument within the medical profession at the time of this writing is not whether surgery should be required, but whether it should be offered at all.

III. THE EVOLUTION OF IDEAS

A. Parental Authority and Limitations on the Right to Refuse

As discussed in the first section,³⁰ the right of a competent adult patient to refuse a medical therapy, even a potentially life-saving one, is now widely accepted in the United States among bioethicists and physicians. Most physicians would, in most settings, make every attempt to persuade the patient if it was felt that his or her life was at risk. Ultimately, however, the patient's right to refuse, and the physician's obligation to respect the patient's autonomy, is generally felt to trump other ethical concerns. It is a fundamental misunderstanding, however, to believe that this same right extends to parents who refuse such treatments on their child's behalf.

One sometimes encounters the expression "parental autonomy," which is a misnomer. Autonomy literally means self-rule; therefore

30. See discussion *supra* Part I.A.

one cannot have “self-rule” over someone else, even one’s own child. A more appropriate term is *parental authority*. Parents are generally felt to have far-reaching authority over their children, including the right to refuse certain medical treatments. However, that authority to refuse, though significant, is not as absolute as it is in patient autonomy.

1. *Refusal on Religious Grounds*

A classic example commonly given is the adult Jehovah’s Witness who refuses a life-saving transfusion on religious grounds. Such a refusal is generally respected, and this might be seen as an example of the right to religious freedom. That same individual, however, is generally not accorded the same right to refuse on behalf of her infant. Neonatologists would generally seek to overrule such a parental refusal, and in a situation where there was not adequate time to consult a court before acting, they would generally act to save the infant’s life. To those who express concern that such an action denies parents their right to freedom of religion, the opinion of the U.S. Supreme Court in 1944 is often cited: “Parents may be free to become martyrs themselves. But it does not follow that they are free, in identical circumstances, to make martyrs of their children”³¹

The AAP has issued a very clear statement in support of such limits on parental authority. In its statement on religious objections to medical care, the AAP Committee on Bioethics states that “physicians who believe that parental religious convictions interfere with appropriate medical care that is likely to prevent substantial harm or suffering or death should request court authorization to override parental authority or, under circumstances involving an imminent threat to a child’s life, intervene over parental objections.”³² The statement goes on to say, however, that “when caring for children whose prognoses are grave even with treatment,

31. *Prince v. Massachusetts*, 321 U.S. 158, 170 (1944).

32. American Academy of Pediatrics Committee on Bioethics, *Religious Objections to Medical Care*, 99 PEDIATRICS 279, 280 (1997).

physicians should use restraint in pursuing a court order to initiate or continue treatment when parents object to it.”³³

Thus, with regard to religious objections, it would appear that the AAP reasonably suggests that there is some threshold beyond which parents no longer have a right to refuse. That threshold is defined by the phrase “likely to prevent substantial harm or suffering or death.”³⁴ If the infant is very likely to die regardless of treatment, then overruling the parental refusal is not appropriate. A specific example with regard to transfusion of an extremely premature newborn has been discussed in the medical ethics literature.³⁵ However, the wording and intent of the AAP is quite clear that parents’ right to refuse treatment stops when the above threshold has been met.

2. Generalization of the Limit to Parental Authority

The restriction on parental authority cited above, and supported by both the *Prince v. Massachusetts* ruling and the AAP policy, is not limited to refusal on religious grounds. It is clear that the intent of the AAP statement was to set the threshold cited for any parental refusal. Indeed, in another statement on forgoing life-sustaining medical treatment, the AAP endorses the parents’ right to make the value judgments so inherent to such difficult decisions and advises physicians to respect their choices. However, that advice comes with the following caveat: “Medical professionals should seek to override family wishes only when those views clearly conflict with the interests of the child.”³⁶

It is the best interests of the child, then, that should ultimately guide our decisions regarding withholding treatment. The child’s best interests are generally assessed by weighing the anticipated benefits

33. *Id.*

34. Mark R. Mercurio, *Parental Refusal of Transfusion on Religious Grounds: Exceptions to the Standard Approach*, 2 CLINICAL ETHICS 146, 146 (2007) (quoting American Academy of Pediatrics Committee on Bioethics, *supra* note 32).

35. *Id.* at 146.

36. American Academy of Pediatrics Committee on Bioethics, *Guidelines on Forgoing Life-Sustaining Medical Treatment*, 93 PEDIATRICS 532, 533 (1994).

of the proposed treatment to the child against the burdens to the child. Benefits of a treatment might include a chance of survival and the pleasures that years of living might bring. Burdens might include the pain of treatment and ongoing intensive care, particularly if the chance of survival is low, and the burden of ongoing disability. These will be subjective judgments, and we should generally defer to the judgment of parents until some threshold is reached (in other words, “clearly conflict with the interests of the child”).³⁷

The most recent statement issued by the AAP on non-initiation or withdrawal of intensive measures specifically for the case of newborns reiterates the basic premise that treatment decisions should be guided primarily by the best interests of the child: “[t]he physician is not obligated to provide inappropriate treatment or to withhold beneficial treatment at the request of the parents.”³⁸ The policy is clear that withholding or withdrawing treatment is an appropriate decision for some newborns. What is less clear, and where the moral work is found, is determining the degree of burden to the patient, including severity of prognosis and anticipated disability, that justifies a parental refusal of treatment. Few if any American neonatologists today would argue that the anticipated burden of disability to children with Trisomy 21, such as Baby Doe, justifies parental refusal of treatment.

3. *Physician-Parent Disagreements and the Notion of Futility*

In the quarter-century since the Baby Doe Rules, there has been an interesting shift in the pattern of conflicts between parents and physicians about medical treatment. While parental refusal of recommended treatment still occurs, there seems to have been an increasing frequency of parental demands for treatments against the advice of physicians. For the three different hospital ethics committees that have been chaired by this author during the past two

37. *Id.* (emphasis added).

38. American Academy of Pediatrics Committee on Fetus and Newborns, *Noninitiation or Withdrawal of Intensive Care for High-Risk Newborns*, 119 PEDIATRICS 401, 402–03 (2007).

decades, that has clearly been the case. Request for assistance with disagreement over patient or parent demands for inappropriate (in the view of the physicians) treatments far outnumber those for patient or parental refusals. Physicians in general, and neonatologists specifically, have struggled with how to deal with such demands. One response has been to invoke the concept of futility.

The concept of medical futility seems to date from the 1980s and has been a focus of disagreement ever since. Physicians often justify a refusal of parent or patient demands by saying that the requested treatment is futile, and they are therefore not obligated to provide it. The basic notion of futility seems relatively straightforward; it is a common English word meaning fruitless, or unable to accomplish the desired end. The term is not unique to medicine, but nevertheless physicians and ethicists have debated its meaning in the medical context for many years, and have worked to craft policies and guidelines for dealing with family demands in “futile” situations. Its use as a justification of physician refusal has fallen into disfavor for some in recent years, however, perhaps in part because some physicians have used it to avoid difficult conversations about the rationale for withholding further treatment in very difficult (sometimes hopeless) situations.³⁹ Simply by invoking the concept of futility a physician may seek to avoid having to explain exactly why a treatment is not appropriate. Nevertheless, its use in some settings continues, and even the most recent AAP guidelines on intensive care for newborns states that the physician is not obligated to provide inappropriate treatment, and “treatment that is harmful, of no benefit, or futile and merely prolonging dying should be considered inappropriate.”⁴⁰

Whether or not one accepts the use of the term “futile” in the medical context, it seems right that physicians should be able to refuse to provide treatment that they feel cannot accomplish a

39. See Jeffrey P. Burns & Robert D. Truog, *Futility: A Concept in Evolution*, 132 CHEST 1987, 1991 (2007); Helft et al., *The Rise and Fall of the Futility Movement*, 343 NEW ENG. J. MED. 293, (2000).

40. American Academy of Pediatrics Committee on Fetus and Newborns, *supra* note 38, at 402.

worthwhile goal or benefit the patient. The difficulty lies in determining what goals are worthwhile. If parents awaiting a miracle demand ongoing intensive care for a child who seems certain not to survive the hospitalization, is that worthwhile? If parents demand resuscitation of a preterm newborn with an estimated chance of survival of 4%, and of survival without significant disability of 1%, is that worthwhile? If parents demand cardiac surgery so their child with Trisomy 13 can live longer, albeit in a profoundly disabled state, is that worthwhile? One may attempt to apply the patient's Best Interests Standard to these questions, and still find them troubling. Below what threshold of likelihood of survival, or what degree of predicted disability, does the burden of ongoing treatment outweigh the benefit? These are now the questions that plague neonatologists far more than refusal of recommended treatments.

B. Present and Future Issues

In closing it is worth briefly considering some current issues that are likely to influence the ethical debates in neonatology in the near future.

1. Consideration of the Interests of the Family

Pediatric ethicists and others have widely endorsed the patient's Best Interests Standard when making decisions regarding life-sustaining treatments in children, including newborns. This is evident in several of the documents referenced throughout this manuscript. This standard requires that such decisions be based on weighing the relative benefits and burdens *to the child* of the proposed treatment. However, the decision to keep a severely disabled newborn alive could have a significant impact on the life not just of that newborn but on the lives of others as well. Duff and Campbell, and others, have suggested that it is acceptable to consider the interests not only of the child but also of other family members when deciding for a newborn patient. This approach may already be at work in some

NICUs.⁴¹ It is beyond the scope of this essay to argue whether the interests of the parents or the siblings should be considered. It is, however, here suggested that an open and frank dialogue of this question is needed, and if it is ultimately determined that such a consideration is appropriate, then policies and guidelines should be rewritten to reflect that view.

2. Consideration of the Interests of Society

If we believe it is fair to consider the interests of people affected by these decisions, it does not necessarily follow that this must be limited to family members. Another logical challenge to the patient's Best Interests Standard would be to consider the interests of society, which will in many cases bear some of the burden of ongoing care for many of these children, often for many years, possibly for decades. Society will bear that burden in a setting of increasingly limited resources, and it will be argued by some that those resources could more wisely (or more fairly) be spent elsewhere. This argument could be brought to bear on some of the examples discussed above, such as the provision of intensive care to an extremely tiny newborn with a 5% chance of survival, who will likely suffer significant impairment if he survives, or the provision of extended intensive care or cardiac surgery to newborns with Trisomy 13 or 18.

Some might see this as a necessary step to provide the most good for the most patients, given limited resources. Others might be concerned that this approach is reminiscent of warnings brought forth by Dr. Alexander six decades ago, and the Nazi concept of some members of society being "useless mouths," and thus not deserving of ongoing care. However, it seems unfair to characterize those who seek a more just distribution of resources in this way. An open, frank, and respectful dialogue about this fundamental question of medical ethics clearly needs to occur.

41. J. HARDWIG, *IS THERE A DUTY TO DIE? AND OTHER ESSAYS IN MEDICAL ETHICS* 29–43 (Routledge 2000).

3. *Euthanasia*

It is now widely accepted by neonatologists and medical ethicists in the U.S. that, in certain settings, withholding life-sustaining treatment from some newborns is acceptable. In some cases this will be the judgment even if it is understood that, by doing so, death will inevitably result within minutes or hours. One might then ask whether it is acceptable to provide that same newborn, by active means, with a quick and painless death. Ethicists have long debated the similarities and differences of killing versus letting die, but at the time of this writing euthanasia is not an openly accepted practice in NICUs in the U.S. There is not, however, universal agreement on this question.

Dr. Alexander's manuscript referenced above made particular note of the heroism of Dutch physicians who, unlike their German counterparts, seemed to successfully resist the attempts by the Nazi regime to make them complicit in acts of euthanasia.⁴² It is an interesting arc of history that the most widely known legal protocol for euthanasia of profoundly damaged newborns, the Groningen protocol, is currently in place in the Netherlands.⁴³ Here again, an open, honest, and frank dialogue seems indicated.

4. *Better Predictive Data*

The Baby Doe case in Bloomington, and the regulations that soon followed, occurred in the wake of the establishment of neonatology as a new certified sub-specialty within pediatrics less than a decade before. The initial court ruling regarding Baby Doe makes no reference to testimony from a neonatologist, and it does not seem that the patient was ever seen by one. In the years that have followed, medical decisions about withholding treatment from newborns have generally been made by neonatologists, or at least with their active involvement. Such decisions (and advice in these settings) should be

42. Alexander. *supra* note 6.

43. A.A. Verhagen and P.J. Sauer, *End-of-Life Decisions in Newborns: An Approach from the Netherlands*, 116 *Pediatrics* 736-39 (2005).

based on the best and most current prognostic data available, as well as cautious and realistic assessment of the accuracy and relevance of those data. The decision in the Baby Doe case might be seen as the application of sound ethics to bad prognostic information (no chance of any quality of life). It is incumbent on neonatologists to seek and use reliable prognostic information when counseling parents and attempting to resolve difficult questions about withholding treatment. The Tyson paper and the Outcome Estimator based on the NICHD database represent major progress in that regard. Much more work needs to be done in other aspects of neonatal care and prognosis. And, where there are no reliable data upon which to rely, physicians need to be honest with parents, and with themselves, that such is the case. Even in the face of a most thoughtful and sophisticated ethical deliberation, bad decisions might be reached if bad data are applied.

