Who Is the Next "Baby Doe?" From Trisomy 21 to Trisomy 13 and 18 and Beyond

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abstract

The "Baby Doe" case of the early 1980s was marked by considerable controversy, primarily regarding the legal response of the federal government to the case at the time. In the decades that followed, the decision-making for children with trisomy 21, like Baby Doe, has been substantially reevaluated. The data, the assumptions about quality of life that were based on those data, and the ethical principles underpinning the decision-making in the Baby Doe case have all evolved significantly over time. The present strategies for decision-making for children with trisomy 13 and 18 appear to be following a similar pattern. The data, quality-of-life assumptions based on those data, and even the ethical principles underlying the decision-making for these children are currently being reexamined. Children with trisomy 13 and 18 are, in this regard, the next Baby Doe(s).

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In Bloomington, Indiana, in 1982, a child was born whose too brief life would have a profound and lasting effect on modern medicine. The controversy that followed his death would be used to lay the foundation for the practice of medical ethics and change the course of pediatric decision-making forever: the concepts of the best interests standard,[1] constrained parental autonomy,[2] and the harm principle[3] would emerge in the decades that followed. But who is the next "Baby Doe?" Who are the children currently being treated whose cases, with time, will cause a similar sea change? Which decisions that seem so clear in the present will look so different from the future? To answer these questions, it is important to first examine key features of the Baby Doe case. In doing so, similarities will emerge between that historic case and our present care of children with trisomy 13 and 18. Children with trisomy 13 and 18 are, in some sense, the next Baby Doe(s) for whom a critical reevaluation of medical practice is underway.

THE HISTORY OF BABY DOE

Baby Doe was born with trisomy 21 as well as esophageal atresia and tracheoesophageal fistula[4] (congenital anomalies that prevented him from eating safely but were correctable with a straightforward surgical procedure). His parents were told by members of the medical team that because of his trisomy 21 he would be severely disabled and would have an unacceptable quality of life.[5] On that basis, they declined what would have been life-saving surgery, and he died in the hospital nursery 6 days later.[4] His case received substantial public attention and sparked the development of the federal Baby Doe regulations requiring hospitals to post hotline numbers in public spaces and authorizing Baby Doe Squads to investigate complaints of medical care being withheld from newborns with disabilities.[5,6] These regulations created a firestorm throughout the medical community,[6,7] and the American Academy of Pediatrics successfully sued the federal government: the first set of Baby Doe regulations were struck down in 1983.[6]

However, although the American Academy of Pediatrics' legal victory was largely viewed as a success, the way that Baby Doe was treated is no longer considered acceptable medical practice. It is no longer ethically permissible to withhold life-saving surgical interventions from children with trisomy 21 in the absence of serious complicating clinical factors (such as severe congenital heart disease).[5,8] This is in large part due to a reevaluation of the clinical data about trisomy 21 and a recognition that quality of life is not nearly as diminished as was believed in the 1980s.[5] The change in the standard of care for children with trisomy 21 over time also reflects an evolution in the ethics of pediatric decision-making. Key decision-making concepts (the best interests standard,[1] constrained parental autonomy,[2] and the harm principle[3]) emerged in the decades following the case and in response to the questions raised by it.[2,9]

CHILDREN WITH TRISOMY 13 AND 18: THE NEXT BABY DOE(S)

The care of children with trisomy 21 like Baby Doe has changed substantially in the last 35 years. A parallel evolution may be occurring in our care of children with trisomy 13 and 18. For these children, historically, "There was a tacit consensus that life-sustaining treatment was not medically indicated,"[8] and, in some institutions, this is still the approach.[8] This is based in part on the low survival rates anticipated for these children: only ~10% will survive beyond 1 year of age.[10] It is also based on concerns about quality of life and the profound neurodevelopmental impairments experienced by these children.[11,12] However, recent data have revealed that outcomes with intervention may be better than previously thought.[13–16] Some parents of children with these conditions have strongly advocated for therapies that had been traditionally withheld.[8,11] The Support Organization for Trisomy 18, 13, and Related Disorders maintains an online database documenting the frequency with which certain procedures are conducted in children with trisomy and, in the case of cardiac surgery, the facilities willing to provide them. The previously clear approach to medical decision-making for these children is being reexamined and with good reason.

EMERGING DATA ON SURVIVAL

In past discussions of children with trisomy 13 and 18, researchers have failed to note that although a low number of children survive to age 1, those who do survive tend to continue to do so.[13] For example, although at birth the likelihood that a child with trisomy 13 will survive 1 year is only 20%, if the child survives 6 months, the chance of survival to 1 year is 77%.[13] If the child survives 1 year, the 10-year survival is 65%.[13] The sharp decline in survival in the early part of the Kaplan-Meier curve for these children also has been shown to raise the possibility of self-fulfilling prophecies.[5,8] Delivery room or neonatal interventions may commonly be withheld or withdrawn from these children on the basis of their low survival. As a result of therapy being withheld or withdrawn, few children survive, and then therapy is withheld or withdrawn from other children on the basis of the observed low rate of survival. However, the critical question is not how many children survive, but how many children would survive if we intervened.[5]

As indicated in a growing body of data, more medical intervention may be done to improve the survival of these children. In one single-center study of children with trisomy 13 who received intensive neonatal and pediatric treatment without cardiac surgery, researchers reported a median survival of 451 days and 1-year survival rate of 54%.[14] Other studies have also revealed improved survival when cardiac surgery was provided.[15,16] One challenge is that these data are largely from population-based observational studies and so suffer from immortal time bias, in which the children who survive long enough to undergo surgical intervention may be fundamentally different from other children in the study.[17] Additionally, there are increased risks and costs associated with these procedures that are not yet fully understood.[18,19]

EMERGING INFORMATION ON QUALITY OF LIFE

In addition to historically low survival rates, the other rationale for withholding life-sustaining therapies from children with trisomy 13 and 18 is concern about neurodisability and their future quality of life. Although it is true that essentially all children with these conditions experience profoundly impaired neurodevelopment, research reveals that children with trisomy 13 and 18 continue to achieve developmental milestones over time.[12,14] This is echoed by what parents have described in the literature.[11] More importantly, intellectual disability (even profound disability) does not necessitate an unacceptable quality of life.[20] In a seminal study of parents of children with trisomy 13 and 18 involved in online communities, the authors state, "Although most parents described their children as having significant neurodevelopmental disabilities, almost all parents reported a positive view of family life and the quality of life of their child with T13-18."[11] In that study, it is stated that "99% of parents described their child as a happy child."[11] These parents described a different picture in their lived experiences than what was described to them by medical professionals.

BABY DOE REDUX

In that we are reevaluating both the data and the meaning of that data for children with trisomy 13 and 18, there are clear parallels with the Baby Doe case of the 1980s. However, there is at least one important difference. In the Baby Doe case, therapy was withheld at the family's request. Today, it is no longer acceptable to withhold life-saving surgery from a child with trisomy 21 (without severe complicating factors), even if the family requests it.[5] Life-sustaining treatment of these children has, for the most part, become mandatory. In cases of trisomy 13 and 18, however, decisions to unilaterally withhold therapy are what is being reexamined. The acceptability of withholding life-sustaining therapy without a family's input (or over their objection) is being reevaluated. Given the severity of trisomy 13 and 18, however, burdensome life-sustaining therapies are unlikely to become mandatory. Even as new data emerge, the treatment of children with trisomy 13 and 18 is likely to remain in the "stable gray zone"[8] in which parental input is paramount.

The Baby Doe case also was used to spark a critical reevaluation of the ethics of decision-making for children, culminating in the development of the principles of the best interests standard, constrained parental autonomy, and the harm principle. The care of children with trisomy 13 and 18 and those with profound neurodisability may be the source of another period of similar ethical evolution. Our attitudes about the value of children who will have a "short, disabled life"[20] and the need for "diversity of lives and choices" is evolving.[20] The importance of relational potential in decision-making for children is being explored.[21] Some (including this author) argue that, when making decisions for children with trisomy 13 and 18 and other conditions of profound neurodisability, the "best interests standard is too limited. John Arras's 'relational potential standard,' conjoined to a contemporary care ethics framework, provides a better guide."[21] As our practice continues to evolve, new ethical concepts and principles are likely to emerge.

SUMMARY

In the 1980s, pediatricians grappled strenuously to come to consensus on the appropriate care of children with trisomy 21. Those decisions seem so obvious in retrospect: the vehement disagreement over them seems curious from this side of the millennium. Which decisions that seem so clear now (or even that are deeply controversial in the present moment) will look different in the future? There has recently been the beginning of a reevaluation the medical care of children with trisomy 13 and 18. As the data are examined and interpreted and new ethical concepts emerge, the care of these children is likely to continue to evolve. One physician writing in *Pediatrics* on the Baby Doe case noted that "Decisions concerning the care of defective newborns are difficult. Physicians and families rarely make these decisions lightly; significant effort and introspection are contributed to these decisions by both parties."[7] She went on to explain that "the method by which ethical decisions are made must be flexible enough to provide the best decision for that infant and family."[7] Some 35 years later, this complex, difficult endeavor remains a work in progress.

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