

Attitudes of Neonatologists Toward Delivery Room Management of Confirmed Trisomy 18: Potential Factors Influencing a Changing Dynamic

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The authors have indicated they have no financial relationships relevant to this article to disclose.

What's Known on This Subject

Trisomy 18 is a common genetic disease with a very poor prognosis that traditionally has been classified as a futile condition, for which resuscitation is not indicated.

What This Study Adds

This study contributes the current responses of neonatologists to the ethical question of resuscitation and intensive care for an infant with confirmed trisomy 18. This article also addresses the change in providers' ethical reasoning away from a best-interest standard.

ABSTRACT

OBJECTIVE. The objective of this study was to determine the attitude of neonatal providers toward delivery room resuscitation of an infant with confirmed trisomy 18 with known congenital heart disease at ≥ 36 weeks of gestation.

METHODS. A multiple-choice questionnaire listing this clinical scenario was completed by neonatologists and fellows staffing level III NICUs. Potential factors influencing the decision to initiate resuscitation included maternal preference, neonatal condition at birth, obstetric care, and legal concerns.

RESULTS. Fifty-four (76%) of 71 surveys were completed. Of respondents, 44% indicated that they would be willing to initiate resuscitation. Maternal preference (70%) was the primary reason to initiate resuscitation, with the appearance of the neonate in the delivery room (46%) and legal concerns (25%) as additional factors.

CONCLUSIONS. Until recently, there was universal consensus that trisomy 18 was a lethal anomaly for which resuscitation in the delivery room was not indicated. These data indicate that more providers (44%) than anticipated would consider initiation of resuscitation for an infant with trisomy 18 even with congenital heart disease. We speculate that support for the best-interest standard for neonates is diminishing in favor of ceding without question to parental autonomy. This shift may have profound implications for ethical decisions in the NICU. *Pediatrics* 2008; 121:1106–1110

www.pediatrics.org/cgi/doi/10.1542/peds.2007-1869

doi:10.1542/peds.2007-1869

Key Words

ethics, neonatal resuscitation, trisomy 18

Accepted for publication Oct 5, 2007

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AMONG THE MANY ethical dilemmas encountered in neonatal medicine, one of the most troubling and contentious relates to whether to offer aggressive resuscitative measures for infants diagnosed as having conditions that carry extremely poor prognoses. Although many articles have explored neonatologists' attitudes toward resuscitation decisions at the gestational edge of viability,¹⁻⁷ where intact survival rates are low, few have focused specifically on the decision-making process for infants with confirmed genetic anomalies such as trisomies 13 and 18. Historically, there was tacit consensus among those providing neonatal intensive care that these were lethal trisomies and therefore were classified as conditions for which resuscitation was not indicated.

It is appropriate and critically important that physicians strive to limit the futile use of neonatal intensive care, because by definition such care causes some degree of pain and suffering without offering any reasonable hope of benefit.⁸ Concurrently, providers should strive to partner with parents in providing care and should try to accede to parents' wishes regarding their newborn as much as possible.^{9,10} These 2 goals of avoiding futile care and honoring parents' wishes have the potential to come into direct conflict during the process of making difficult neonatal ethical decisions. This possible quagmire can be negotiated by appealing to the governing ethical standard for individuals who make decisions for neonates, that is, to act in the best interest of the infant.¹¹ The best-interest standard ultimately should supersede any ethical claim that parents might make under the heading of autonomy or that society might make under the heading of justice or a desire to reduce the burden to the community.^{9,10,12-18} The imperfections and difficulties in the application of this ethical standard have been well documented in the bioethics literature,¹⁶⁻¹⁹ but this has remained the assumptive ethical stance of both parents and pediatricians caring for sick infants, in large part because of its singularity in asking that both parties in this partnership adopt a strictly child-centered stance in all treatment decisions.

TABLE 1 Questionnaire

You are called to the delivery room for the following clinical scenario. Delivery is imminent. Please indicate whether you would consider initiating resuscitation, incorporating the constellation of various factors that affect your decision. The pregnancy is at 36 wk. The fetus has been prenatally diagnosed with trisomy 18 and congenital heart disease.

Would you consider initiating resuscitation? yes/no

If yes, please rank in order from 1 to 3 the factors that would influence you toward resuscitation (with 1 being the greatest driving force)

Personal medical opinion that this condition is compatible with survival

Mother's indication that she wants "everything done"

Obstetric management decisions, such as performing a cesarean section

Heart rate of >100 beats per min in the delivery room before any resuscitative intervention

Neonate appearing "vigorous" at birth

Concern over legal ramifications if resuscitation is not offered

Neonatal intensive care should be viewed as a trial of clinical therapy, with a short-term goal of avoiding morbidity and death and a long-term goal of attaining or preserving a functional state.^{20,21} In each case in which it is considered, the potential benefit (survival or attainment of a functional state) versus the cost of suffering should be evaluated.

For most neonatal patients, there is clear net benefit to intensive care therapy, and physicians are obligated to advocate for treatment even if it is against the wishes of the family.²² For infants with no reasonable hope of benefit, physicians have the same obligation not to offer therapy, regardless of the wishes of the family, because the intervention would be futile.^{8,23,24} In the latter circumstances, offering therapy despite the knowledge of futility would be a violation of the ethical principle of non-maleficence (do no harm). Admittedly, futility is an ethical concept fraught with the potential for abuse as a self-fulfilling prophecy; therefore, its injunction must be used carefully. Therapies that offer no reasonable chance of bringing about the desired short- and long-term goals of care are correctly classified as futile. Under circumstances in which there is physician-parent conflict regarding the patient's best interest in the aforementioned situations, a third party, such as an ethics committee, may be needed to help find a resolution.

There is a third category in the decision-making matrix regarding whether to treat, which is often referred to as the "gray zone" and is characterized by uncertainty regarding what is in the infant's best interest. Parental wishes should be the deciding factor under such circumstances, if the weight of medical thought cannot firmly establish a consensus regarding the patient's best interest.^{14,25} The new international neonatal resuscitation guidelines highlight the importance of incorporation of parental wishes by stating that parents would like to be given a greater voice in decision-making.²⁶ However, it is also stated in those guidelines that this must be counterbalanced against futile conditions associated with almost-certain death or an unacceptably high morbidity rate among rare survivors, such as cases of confirmed trisomy 13 or 18, for which resuscitation is not indicated.²⁶ Interestingly, the most-recent American Academy of Pediatrics neonatal resuscitation guidelines omit trisomy 18 from the list of examples of conditions for which resuscitation is not indicated.²⁷ The reasoning behind this change from previous guidelines is not entirely

clear. To investigate whether there was indeed a subtle shift in attitude on this topic, we were interested in determining the current approach of delivery room providers regarding resuscitation decisions for infants with confirmed trisomy 18. The objective of this study was to determine whether providers would be willing to consider resuscitation of an infant with confirmed trisomy 18 and congenital heart disease and, if so, to determine which factors were most likely to influence them toward a decision to resuscitate.

METHODS

A multiple-choice questionnaire was mailed to neonatal fellows and attending neonatologists in level III NICUs in the greater metropolitan area of New York City. The units¹² were chosen largely from the referral network of the authors' institution and did not represent an exhaustive list of units in the New York City area. Questionnaires were mailed directly to the providers at their place of work. A self-addressed, stamped envelope to return the survey was included. There were 2 mailings, with identical recipient lists, separated by ~1 month.

In the questionnaire, providers were presented with the scenario of being called to the delivery room for the delivery of a 36-week fetus with confirmed trisomy 18 and congenital heart disease (Table 1). The first question regarded whether the provider would consider resuscitation (yes or no). If the provider indicated that he or she would consider resuscitation, then the respondent was then asked to identify which factors would most influence his or her intervention toward initiation of resuscitation. Respondents could choose up to 3 reasons, ranked 1 (most significant) to 3. Each reason to initiate resuscitation was counted as a positive response if the respondent scored it 1 or 2. The only identifying information in the survey was the gender of the provider and the number of years he or she had been practicing neonatal medicine (presented as a multiple-choice range of years).

The data were analyzed by using χ^2 or *t* tests where appropriate. The questionnaire was approved by the institutional review board of Weill Cornell Medical College.

RESULTS

Seventy-one surveys were mailed, and 54 surveys (76%) were returned completed after 2 mailings. There were 29 male respondents and 25 female respondents. Twenty-four

(44%) of 54 respondents indicated that they would consider initiation of resuscitation of an infant with trisomy 18 and known congenital heart disease.

There was no difference between male and female respondents and no influence in terms of years in practice. Maternal preference was the primary reason to consider initiation of resuscitation, noted by 22 (92%) of 24 of respondents. The condition of the neonate in the delivery room (appearing vigorous or having a heart rate of ≥ 100 beats per minute) was the second most cited reason (11 of 24 respondents; 46%), with legal concerns as the third factor (6 of 24 respondents; 25%). A small minority of respondents (3 of 24 respondents; 11%) chose previous obstetric management decisions or personal medical belief that this was a condition compatible with survival.

DISCUSSION

Trisomy 18 is the second most common triploidy after trisomy 21, affecting ~ 1 infant per 8000 live-born infants.^{28,29} The median life expectancy for an infant with nonmosaic trisomy 18 who is not treated with intensive care is quoted as 2 to 10 days.^{28,30–33} Survival statistics indicate that first-year mortality rates are between 90% and 100%,^{28,32,33} and the vast majority of these infants die in the first month. Those who do live beyond the first year experience shortened lives marked by severe neurologic and physical impairment.^{34,35}

Even with the support of full intensive care, the data indicate that infants with trisomy 18 do not flourish. Two recent international publications detailed the “natural history” of infants with trisomy 18 who receive intensive care, including intubation and surgery. A Japanese study ($n = 24$) reported a 1-year survival rate of 25% and a median survival time of 152 days.³⁶ A Polish study ($n = 20$) found that, with full intensive care support, 30% of the infants survived to discharge from the NICU. Although the authors acknowledged that the impact of widespread aggressive treatment on mortality rates is unclear, they raised the concern that, even with the most aggressive care, most infants in their study died before discharge from intensive care; the authors urged the avoidance of invasive procedures in light of the poor prognosis.³⁷

The hypothetical case in this questionnaire was complicated by congenital heart disease, with a likely need for corrective heart surgery. In one of the few review articles on the subject, Paris et al⁸ clearly presented an ethical argument that surgery is not in the best interest of these infants. This position of nonintervention was reaffirmed in the most-recent recommendations from the Neonatal International Liaison Cardiopulmonary Committee on Resuscitation, in terms of delivery room resuscitation.^{26,27} The conclusion that intensive care is inappropriate for these infants was also supported in the ruling in a British court case.³⁸ In that case, a family court judge ruled in favor of physicians who had been sued by a mother for their refusal to initiate mechanical ventilation for her newly born son with trisomy 18. The judge explained her ruling in terms that reflected the best-interest standard. She reasoned that, if the infant was placed on a ventilator, then he would necessarily be

deprived of activities that certainly were in his best interest, such as being able to be in his mother’s arms, and the judge decided that such deprivation would be ethically and legally unacceptable. This case is particularly interesting because the judge was clearly motivated to determine which specific features of life were beneficial to the infant and to preserve those elements.

Until recently, the medical community that cared for infants with trisomy 18 seemed to agree that it did not serve these infants’ best interests to undergo invasive procedures, such as heart surgery. A commentary on the issue has challenged this premise,³⁹ and the findings in this report suggest that there is a changing approach, largely driven by physicians’ desires to honor parents’ preferences. The vast majority of respondents ($\sim 90\%$) who would consider resuscitation indicated that they would do so despite knowing that the life span of infants with trisomy 18 is invariably short. These observations raise the concern that some neonatologists are abandoning the best-interest standard, which would require that providers agree only to treatment strategies that are consistent with furthering the good of the infant, and instead are adopting an “ethic of abdication” in their approach to difficult treatment/non-treatment decisions.⁴⁰ A significant proportion of providers seem to be willing to put aside professional responsibility to direct the treatment decision and instead concede to any decision that parents may make. This may be a byproduct of the strong emphasis bioethics has placed on patient autonomy and the extension of this concept to “parental autonomy” in decision-making for children.³⁹

In contrast to the relative silence on this topic in the medical literature, in recent years there has been an increase in Internet sites devoted to the most common fatal trisomies (trisomies 13 and 18) and aimed at parents who have received these devastating diagnoses. These sites offer both a wealth of helpful information and a ready-made support community in a time of true crisis and, in that regard, they provide an invaluable service. However, the authors of those sites are assumptive in their opinion that all medical options should be offered to parents, from the moment of prenatal diagnosis to the delivery room and beyond. Resuscitation, intensive care, and surgical corrections are presented as equally valid alternatives to comfort care.⁴¹ For parents who have been informed of this diagnosis prenatally and are searching for all information they can gather, expectations that full resuscitation is a reasonable approach are developed. These expectations are then transmitted to both the obstetrician and the neonatologist and may explain in part the willingness of such a large proportion of respondents in this study to consider resuscitation.

Neonatologists should strive to promote bonding between parents and infants and have a moral duty to care for and to enrich each brief life to the extent possible. Infants with lethal congenital anomalies and profound neurologic impairment continue to have immense inherent worth as human beings, and these infants are without doubt as deserving of love, care, and dignity as any child. However, we contend that having intensive care measures such as intubation and corrective surgery available as potential options for infants with a con-

firmed lethal trisomy gives the impression to parents that these are reasonable interventions to consider and downplays the invasive painful nature of these therapies. By choosing this nonguiding approach, physicians may place undue guilt on parents who would choose comfort care only, because implicit in the latter decision is the understanding that the parents could have chosen to do more. If we as physicians could be clear that noninvasive support and comfort care are the appropriate therapies for some infants, then some of the gut-wrenching struggle that parents face under such extraordinarily difficult circumstances could be alleviated.⁸ We would also be preserving our fiduciary ethical responsibility to both the infant and the family.

CONCLUSIONS

In this era of rapid access to sometimes-incomplete medical information, communication between the physicians and the parents must occur early and often. In the context of the ethical dilemma discussed above, we suggest that the neonatologist, in conjunction with the obstetrician and a genetics counselor, meet with the parents soon after the prenatal diagnosis is made. The medical team should explain compassionately the futility of any intensive interventions in meeting long-term goals of care, and the plan for palliative care should be delineated clearly. The uncertainty in terms of expected length of survival should be explained; many infants die within the first few days of life, but some live for months and may be able to be discharged from the hospital with the family, with very limited neurologic status. It is critical that the emphasis should remain on fostering the best interest of the infant at every branch of the treatment decision tree. In this way, the family and the medical team can act in concert to reach the shared goals of avoiding hopeless suffering and maximizing all good that can be attained during the infant's short but precious life.

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STUDY FINDS MANY PATIENTS DISSATISFIED WITH HOSPITALS

“Washington—Many hospital patients are dissatisfied with some aspects of their care and might not recommend their hospitals to friends and relatives, the federal government said Friday as it issued ratings for most of the nation’s hospitals, based on the first uniform national survey of patients. The survey was meant to provide a constructive way for patients to complain about arrogant doctors, crabby nurses and dirty or noisy hospital rooms. Medical experts said that some of the complaints bore directly on the quality of care. Many patients reported that they had not been treated with courtesy and respect by doctors and nurses; that they had not received adequate pain medication after surgery; and that they did not understand the instructions they received when discharged from the hospital. Nationwide, in the average hospital, 67 percent of patients said they would definitely recommend the institution where they had been treated to friends and relatives. Sixty-three percent gave their hospitals a score of 9 or 10 on a scale of 0 to 10. At the average hospital, more than 25 percent of patients said nurses had not always communicated well with them. The new data, part of a survey of patient experiences and perceptions of hospital care, is posted at a government Web site: www.hospitalcompare.hhs.gov. The results provide cause for concern, said Dr Carolyn M. Clancy, director of the Agency for Healthcare Research and Quality, a unit of the Public Health Service. ‘Poor communication is a major source of medical errors,’ Dr Clancy said. ‘If doctors are not listening carefully, patients may not bring up important information. Patients who do not understand discharge instructions are more likely to be readmitted to the hospital or end up in the emergency room.’ Nancy E. Foster, a vice president of the American Hospital Association, agreed that many hospitals needed to do a better job of controlling pain and communicating with patients. Pain control keeps patients comfortable and can speed healing and reduce complications after surgery. Many large teaching hospitals scored below the national average on questions about the cleanliness and quietness of the hospital environment. The data came from questionnaires completed by a random sample of patients treated at more than 2,500 hospitals from October 2006 to June 2007. Some hospitals chose not to cooperate, but they will soon have a financial incentive to do so. Herb B. Kuhn, a Medicare official, said that if hospitals did not report the data, the Medicare payments could be reduced, by about \$100 for a typical case.”

Pearl R. *New York Times*. March 29, 2008

Noted by JFL, MD