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Using Patient-Centered Care After a Prenatal Diagnosis of Trisomy 18 or Trisomy 13
A Review

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IMPORTANCE Patient-centered care (PCC) has been advocated by the Institute of Medicine to improve health care in the United States. Four concepts of PCC align with clinical ethics principles and are associated with enhanced patient/parent satisfaction. These concepts are dignity and respect, information sharing, participation, and collaboration. The objective of this article is to use the PCC approach as a framework for an extensive literature review evaluating the current status of counseling regarding prenatal diagnosis of trisomy 18 (T18) or trisomy 13 (T13) and to advocate PCC in the care of these infants.

OBSERVATIONS Extensive availability of prenatal screening and diagnostic testing has led to increased detection of chromosomal anomalies early in pregnancy. After diagnosis of T18 or T13, counseling and care have traditionally been based on assumptions that these aneuploidies are lethal or associated with poor quality of life, a view that is now being challenged. Recent evidence suggests that there is variability in outcomes that may be improved by postnatal interventions, and that quality-of-life assumptions are subjective. Parental advocacy for their infant's best interest mimics this variability as requests for resuscitation, neonatal intensive care, and surgical intervention are becoming more frequent.

CONCLUSIONS AND RELEVANCE With new knowledge and increased parental advocacy, physicians face ethical decisions in formulating recommendations including interruption vs continuation of pregnancy, interventions to prolong life, and choices to offer medical or surgical procedures. We advocate a PCC approach, which has the potential to reduce harm when inadequate care and counseling strategies create conflicting values and uncertain outcomes between parents and caregivers in the treatment of infants with T18 and T13.

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Patient satisfaction with health care is an increasingly significant concern. In 2001, the Institute of Medicine introduced a comprehensive strategy to improve health care in the United States. A key aim of this proposal is patient-centered care (PCC), defined as "providing care that is respectful of and responsive to individual patient preferences, needs, and values and ensuring that patient values guide all clinical decisions."[1(p6)] Based on a comprehensive analysis of patient focus group data, 4 core concepts (dignity and respect, information sharing, participation, and collaboration) have been established to define quality in health care delivery (**Table**).[2] These concepts harmonize with the clinical ethical principles of beneficence, nonmaleficence, justice, and respect for autonomy.[3]

Previous reports suggest dissatisfaction among patient families and growing debate among clinicians regarding care after trisomy 18 (T18) or trisomy 13 (T13) diagnosis.[4,5] Our objective was to use PCC concepts to develop a framework that more fully evaluates sources of dissatisfaction and debate regarding shared decision making and clinical care of infants with T18 and T13. We outline key recommendations for a balanced approach to joint decision making regarding care that has application in other areas of challenging patient treatment.

Background

Trisomy 18 and T13 are chromosomal aneuploidies first identified in the 1960s. These syndromes have high mortality and morbidity and are characterized by growth deficiency, cognitive disability, psychomotor disability, and recognizable patterns of physical anomalies.[6-8] Together, they affect 1 in 1800 US pregnancies; thus, approximately 2000 women will carry a fetus to term with T18 or T13 annually.[8,9]

Trisomy 18 and T13 were historically designated as lethal or incompatible with life[10] and families had traditionally been counseled from this perspective.[6,11,12] Approximately 1:6000 to 1:8000 live births are complicated by T18 and 1:10 000 to 1:20 000 by T13.[6,13] There is increasing evidence of variable outcomes of these infants that may be improved by postnatal interventions.[12,14-17]

Lantos[18] recently addressed the history and evolution of management regarding infants with trisomy 21 that sheds light on the current evolving approach toward infants with T18 and T13. Recommendations to limit treatment for infants with trisomy 21 were previously commonplace; however, it is now impermissible to withhold surgery in such cases (with rare exceptions in complicated circumstances). Lantos explains that ethical decisions regarding treatment futility must include consideration of "survival, neurocognitive deficits, and the burdens of treatment."[18(p397)] He goes on to emphasize that trisomy 18 and 13 once belonged in the category that recommended limited treatment.

There is also increasing debate about quality-of-life (QOL) assumptions.[19-23] Quality of life is subjective and physicians rarely understand the criteria that family use in determining their perception or what the infant might experience. The term *quality of life* is often misused for perceived physical or neurologic impairments; however, some infants can have severe impairments and still have an excellent QOL.[24] Nelson and colleagues[25] recently described improved survival with surgery (primarily cardiac) in a cohort of 428 infants with T18 or T13 over a 21-year period. The median survival time of the 254 children with T18 was 9 days, and the median survival time of the 174 infants with T13 was 12.5 days.[25] Of infants with T18, 13.8% underwent surgery with a 1-year survival rate of 68.6%, and, of infants with T13, 23.6% underwent surgery with a 1-year survival of 70.7%. These results provide a rationale for reconsidering previous recommendations regarding care limitations for these infants. However, Graham[26] questions whether these aggressive interventions have improved survival or the quality of life while using resources at increased cost.

Discussion

Dignity and Respect

Dignity and respect in PCC hold that health care professionals should honor patient perspectives, values, and decisions when implementing care.[27] There is a paucity of literature addressing the influence of particular values and cultural heritage within families on decision making and outcomes after diagnosis of T18 and T13.[28] Physician attitudes accrue from scientific knowledge and experience, yet are influenced by ethical principles and personal beliefs. Patient perspectives are strongly influenced by societal, familial, religious, and cultural factors. The often-required immediacy of medical decision making can make inquiry into a family's values and beliefs clinically impractical. Given the strong spiritual and cultural beliefs that encompass birth and death, both of which may be imminent after diagnosis in T18 or T13, decision making may be difficult. In such situations, individuals have a heightened need for information relevant to their values.

Advances in genetic testing, especially noninvasive prenatal testing and chorionic villus sampling, have led to diagnosis as early as the first trimester of pregnancy. A study of the natural history of fetal T18 found high rates of fetal demise and stillbirth.[29] Pregnancy termination may be offered after T18 or T13 diagnosis; however, availability may be affected by gestational age limitations or cultural confines. The difference between the number of pregnancies affected by T18 or T13 and the live birth rate suggests that fetal demise or pregnancy termination is common. Few studies have assessed cultural differences in attitudes toward prenatal testing and pregnancy termination for broad categories of fetal anomalies,[30,31] and there is inadequate literature addressing the needs or preferences of women who choose this option.

There has been increased acknowledgment of patient preferences in the care of women who choose to continue pregnancies affected by severe or lethal fetal anomalies. The concepts of perinatal palliative care and prenatal advanced birth care planning have been evolving since the theory of perinatal hospice was introduced.[32-40] Wool and colleagues[41-43] published research in developing quality indicators in perinatal palliative care. Perinatal palliative care initiatives are now increasingly supported by clinicians, parents, and professional entities.[21,44] Perinatal palliative care that focuses on interdisciplinary team and family involvement offers a

Table. Application of Patient-Centered Care Approach

Characteristic	Approach
Dignity and respect	•Promote research and dialogue about the influence of patient values, beliefs, and culture on decisions and outcomes after a prenatal diagnosis of trisomy 18 or trisomy 13[4,12,17] •Assess and communicate patient values, beliefs, and preferences at diagnosis and throughout the continuum of care[21,45,46] •Support parents in making decisions that fit with their values[8,15,27,67] •Defer to patient wishes when the prognosis or best interest of the child is unclear[8,20] •Support the development of best practice models and guidelines for perinatal palliative care[45,46] •Create opportunities for providers to listen to the health care experiences of patients with trisomy 18 or trisomy 13 pregnancies or children[2,12,17,41]
Information sharing	•Present accurate figures for survival and outcome that take into consideration the individual clinical features of the fetus or child[8,24] •Avoid the unmodified use of *lethal*, *fatal*, or *incompatible with life* in describing potential outcomes,[8] particularly during pregnancy when the condition and prognosis of the fetus are ambiguous •Provide information about a variety of educational resources, including internet websites and support groups[45] •Consider offering "a positive viewpoint" and "erring on the side of life" within the framework of the Convention of the Rights of the Child and Convention on the Rights of Persons with Disabilities[61] •Advocate a willingness to do whatever it takes to fully inform and understand the perspectives of patients/parents[8,15]
Participation	•Avoid coercion and use good communication to resolve conflicts between patients and health care professionals about pregnancy decisions; arrange for second or third opinions, if necessary, and ethics or legal consultations; allow parents time to consider options on "their own turf" with time-limited goals[3] •Reduce variability in physician approaches to intervention/nonintervention to prolong fetal viability in pregnancy[59] •Petition professional organizations to establish ethical, patient-centered guidelines for care of women with pregnancies complicated by "severe, not uniformly lethal anomalies"[4,17]
Collaboration	•Create interdisciplinary teams that can assess and care for the complex needs of patients/parents throughout the continuum of care[21,36,45,46] •Arrange for ethics consultation for professionals and families with ethical concerns about newborn care to identify potential solutions to conflicts[8,18,22,49] •Establish partnerships with specialist providers and institutions that offer broader options or resources for prenatal or neonatal care, and with agencies that offer ongoing care or support for patients and families[18] •Promote the establishment of centers of excellence for pregnancies complicated by "severe, not uniformly lethal anomalies," and for care of newborns and children with these conditions[12,17]

valuable framework for providing PCC after T18 or T13 diagnosis. Merritt et al[45] provide guidelines for perinatal palliative care in this context. English and Hessler,[46] Wool and Dudek,[47] and Wool[48] offer guidance in outlining steps for assessing patient perspectives and preferences, a shared decision-making process for palliative care, components of a written birth plan, roles of interdisciplinary team members, perspectives of perinatal palliative care barriers, and other valuable guides.

Typical perinatal palliative care after diagnosis of T18 or T13 has 2 important limitations. First, it generally offers limited services to women who opt for pregnancy termination, and second, the common emphasis is on comfort care (which assumes poor neonatal outcome) or providing a compassionate environment to adjust expectations and choose to limit interventions.[45,49] This approach may be hollow if not aligned with patient preferences. van de Eijk et al[50] propose that PCC requires more than just a respectful attitude or a personalized approach to assessment and care; rather, it requires engaging patients to become active participants and decision makers.

Influential parent support groups, such as the Support Organization for Trisomy 18, 13 and Related Disorders (SOFT)[12] and the Trisomy 18 Foundation,[17] have had a significant effect on the dignity and respect afforded patients and families affected by these syndromes. The parent-professional collaborations resulting from the advocacy of parent support groups have vocally advocated over the last decade owing to increased visibility via the internet and additional information in medical and ethical literature. These websites offer contextual information to promote respect for patients and families and provide testimonials of adverse medical experiences while offering videos of children who have overcome the odds of perinatal mortality. Such websites appear, however, to spark controversy among health care professionals, who claim that the sites may promote unrealistic expectations about outcomes due to selection bias toward children who have generally survived for longer than 1 year. However, a survey by Janvier et al[51] of parents of children with T18 or T13 recruited from these social networks emphasizes that parent goals were "to meet their child, be discharged home, and be a family."

Information Sharing

Information sharing requires health care professionals to offer accurate, complete, and unbiased information to patients so that they may make well-informed decisions even when there is controversy regarding management strategies.[52,53] Although patients do not universally appreciate patient-centered information sharing, the ethical principles of respect for autonomy, beneficence, and nonmaleficence support the importance of accurate information disclosure.

Parents are commonly counseled from the perspective that T18 and T13 are "incompatible with life."[54] Population-based studies indicate that mean survival time is between 3 and 14.5 days and 1-year survival is 0 to 10%.[13,55] The length of survival time decreased after T18 and T13 were identified as discrete syndromes, possibly as a result of less aggressive treatment being offered owing to expectation of death.[56] Although recent studies show that mortality remains high, there is increasing evidence of outcome variability and mortality reduction through postnatal interventions. Rasmussen et al[54] agree that aggressive intervention, such as cardiac surgery, may be necessary to prolong the lives of some infants; however, long-term T18 survivors may not need particularly aggressive or extraordinary treatment. According to Niedrist et al, "if children with trisomy 18 are brought beyond the first critical phase of life when postnatal adaptation of cardiorespiratory function is still very poor, the subsequent survival chance is enhanced,"[55(p957)] and the recent report by Nelson et al[25] supports this concept.

Recent studies on T18 and T13 outcomes acknowledge that these aneuploidies are not universally lethal. In a review of genetic screening challenges, Coughlin[57] evaluated surveys of genetic professionals in an attempt to rank the seriousness of genetic disorders. The study confirmed a consensus about which conditions were considered lethal, serious but not lethal, or not serious.[58] In a survey among members of the Society for Maternal-Fetal Medicine, Heuser et al[59] distinguished T18 and T13 as severe, commonly lethal anomalies as opposed to uniformly lethal anomalies, such as anencephaly and bilateral renal agenesis. Despite such variability, specialists routinely use the term *lethal* in counseling the parents of fetuses or infants with T18 or T13 and perpetuate QOL assumptions that promote withholding interventions.[48]

In an international survey of 272 parents of children with T18 or T13 (76% within the United States), parents reported being told by a health care professional that their child's condition was incompatible with life (87%), the child would have a life of suffering (57%), or that care of the child would ruin their family (23%).[4] Of the respondents, more than 25% had a child with T18 or T13 still living, with a median age of 4 years. Although 50% reported that care of a disabled child was more difficult than expected, 97% reported that their child was happy. These study results may be limited by selection bias because the survey did not include women who terminated the pregnancy or experienced fetal loss, or those whose infants died shortly after birth. Parents responding to the survey were identified through T18 and T13 support groups and so may have been more likely to have a living child, be happier with the current status of their child and family, or be more dissatisfied with the information that they received because it proved to be inconsistent with their child's outcomes. Follow-up studies among parents who are not members of support groups may offer a broader perspective on parental perceptions. Janvier et al[4] theorize that increasing disparities in patient and provider perspectives may be due to the aforementioned internet support groups. These testimonials challenge the incompatible-with-life and life-of-suffering perspectives. Mercurio and coworkers[60] observed that physician arguments for withholding treatment have lost credibility owing to insufficient corroborating evidence.

Current approaches to information sharing after diagnosis of T18 and T13 may not be as accurate, complete, and unbiased as they should be.[60] One factor in these limitations may be pressure to make decisions about pregnancy termination before narrow gestational age time frames expire. McGraw and Perlman[19] suggest that parents exposed to support group websites may develop expectations that interventions to prolong life are "reasonable." The context of the comment suggests that physicians may consider parental expectations of prolonging life to be unreasonable. The American Congress of Obstetricians and Gynecologists explicitly states, "it is unethical for a physician to deny patients important information in order to avoid physician-patient interactions that are difficult or uncomfortable."[23(p1025)] Religious, family, and cultural beliefs often influence parental decisions. These decisions are not merely of

weighing potential risks vs benefits; they may, in fact, represent significant spiritual or ethical dilemmas. We advocate a more balanced approach to information shared in the counseling process, including use of up-to-date survival figures taking into consideration individual clinical findings of the child. This approach should include potential surgical interventions and avoidance of the terms *lethal* or *poor quality of life* without presupposition of the family's perceptions. Health care professionals have an obligation to "present prognostic information in a frank and balanced way without coercion."[21(p402)] As proposed by Bruns,[61] they also have an obligation to offer "a positive viewpoint" and "err on the side of life" within the framework of the Convention of the Rights of the Child and Convention on the Rights of Persons with Disabilities.[16,61,62]

Participation

This concept of PCC includes patients' ability to participate in and make decisions about their care at the level they choose.[63] Heuser et al[59] showed that, for women wishing to continue the pregnancy, 99% of physicians would adhere to a patient's wish not to intervene on the fetus's behalf. Five percent would discourage nonintervention, but would adhere to the patient's decision. In contrast, 82% of physicians would adhere to the patient's wish to intervene to prolong the fetus's life (eg, by cesarean section) and 66% would discourage intervention, but adhere to the patient's decision. The perspective appears to be that intervention puts the mother at risk with no clear benefit to a fetus with severe anomalies.

The variability noted in physician approaches raises the ethical principle of justice. Distributive justice assumes that patients with similar conditions are treated alike.[64] In discussions regarding inequity in treatment, the lives of both the fetus with T18 or T13 and the mother may be at stake. Injustice in prenatal management of T18 and T13 may go beyond individual variation in provider approaches to cesarean section. It also involves the broader question of how much personal risk a woman should be allowed to assume to prolong the life of her child. Individuals are often allowed or even encouraged to assume personal risk, such as living organ transplantation, to increase the length or QOL of a seriously ill family member or stranger with no guarantee of positive outcomes. Denying this opportunity for women who wish to assume a similar risk to ensure the live birth of a child with an uncertain prognosis could be considered unjust to both fetus and mother.

If it is known that nonintervention is likely to result in fetal morbidity or mortality, failure to intervene may violate the ethical principle of beneficence, particularly when the mother is requesting the intervention. Although physician approaches vary widely to maternal requests for late trimester pregnancy termination or nonaggressive intrapartum management, Spinnato et al suggest that physicians are obligated to respect maternal autonomy. Spinnato et al[65] explain, "When a patient's desire to avoid an intrapartum stillbirth is strong enough that substantial psychological harm might result from one, the physician's beneficence-based obligation to her and respect for maternal autonomy justify selectively aggressive intrapartum therapy, even if no beneficence-based obligation to the fetus exists."[65(p89)]

Collaboration

Patient-centered care collaboration focuses on involvement of all health care constituents in the development of health care policies.[63] After the birth of a child with T18 or T13, the role of the mother shifts from participant to collaborator in conjunction with other family members and health care professionals. Perceived collaboration in decision making has been described as the most important determinant of parental satisfaction with end-of-life care.[66] Too often collaboration between physicians and parents is compromised by contention. In a survey of 54 pediatricians, 44% indicated that they would be willing to make resuscitative efforts on behalf of an infant with T18 who has known congenital heart disease (present in >90% of T18 cases).[19] The authors characterize the pediatricians' willingness to intervene as adoption of an "'ethic of abdication' in their approach to difficult treatment/nontreatment decisions" as a result of the "strong emphasis bioethics has placed on patient autonomy."[19(p1108)] Physicians frequently center their perception on the best interest of the child, futility of treatment, and waste of resources. Resistance of pediatric specialists to intervene is apparent in a survey of 859 physicians regarding their attitudes toward cardiac palliations in infants with T18 or T13. Neonatologists were least likely (7%) to recommend intervention on heart lesions compared with geneticists (20%) and cardiologists (32%); however, there was a 3-fold increase in willingness to intervene among all specialists if intervention is requested by parents.[53]

Disparate perspectives between parents and clinicians on the best interest of the child, futility of treatment, and allocation of resources may create tension and set the stage for adversarial relationships. Janvier and colleagues[67] illustrate the deep divisions between parents and clinicians caring for infants with T18 or T13. In this case, the parents wanted maximal intervention for their 1-year-old child with T18, including cardiac surgery, and were unmoved by discussions about poor outcomes, risks of surgery, and futility of treatment. It may be difficult for parents to accept that nonintervention is in the best interest of the child when it is likely to hasten the child's death. Finding consensus with parents about futility of treatment may be even more problematic, as medical futility is a poorly defined, highly subjective, and conflict-ridden concept.[68] In the case cited above, the child did well after surgery, which suggests that the parents' request was reasonable and not futile. The variability in outcomes and intensity of interventions for infants with T18 and T13 suggests that it may be difficult to clearly establish the futility of treatment.

In considering allocation of resources, Janvier et al emphasize that "the principle of justice demands similar patients be treated similarly."[67(p758)] They assert that, among other classes of patients who are expected to die or have severe neurodevelopmental compromise, such as those with brain trauma, the general standard is to defer to the wishes of the patient's family. The authors argue that it is unjust to impose restrictions specifically for children with T18 (and, by extension, T13) that are nonexistent for other patients with similarly severe limitations. Furthermore, concerns about wasting resources may be dispelled by the low incidence of live T18 or T13 births and the infrequency of parental requests for maximal intervention.[7]

Collaboration among health care professionals is a particularly important consideration in providing care after diagnosis of T18 or T13. The interdisciplinary structure of models such as perinatal palliative care can provide valuable support for the complex needs of patients and families, particularly if the scope of care is broadened to address the needs of women who choose termination of preg-

nancy. Such teams may be composed of neonatologists, geneticists, palliative care specialists, social workers, ethicists, and nurses—all of whom should have sufficient collective expertise to address the physical, psychosocial, practical, and spiritual needs of the child and family.[21] An ethics consultation may be valuable in cases unsettled by perinatal and neonatal management teams.[21] Such consultations offer a forum for exploration of the family's values and the moral context in which decisions are being made and may result in consideration of a wider range of satisfactory options for care. Lantos suggests that "yesterday's moral gray zones disappear as more data elucidate that treatment is either clearly beneficial or ineffective."[18(p397)] He emphasizes that decisions about aggressive treatment of infants with T18 and T13 are in a "'stable gray zone' in which treatment decisions are unlikely to become more clear"[18(p397)] and align with a shared decision-making approach.[69]

Conclusions

Patient-centered care facilitates decision making after diagnosis of T18 or T13 and other perinatal conditions in which there are conflicting values and uncertainty of outcomes based on patterns of variability of care. Patient-centered care focuses on the parent and child's best interest,[70] offers an ethically sound approach that considers the well-being of all involved, and can be applied in cases of T18, T13, or other conditions requiring complex perinatal decisions.

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