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Moving toward a shared process: The impact of parent experiences on perinatal palliative care

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ABSTRACT

Perinatal palliative care programs seek to support parents expecting a baby diagnosed with a serious medical condition. Clinicians have increasingly recognized the importance of parental perspectives on the medical care mothers and their fetuses and live-born children receive, especially regarding factors influencing individual choices and knowledge of the medical community. We describe, using literature on trisomy 13 and trisomy 18, how information shared between parents and providers can improve perinatal counseling and family support.

Introduction

Every year across the United States thousands of pregnant women receive shattering news from obstetrical clinicians. These women learn through a variety of screening and diagnostic studies that the babies they are expecting suffer from a serious medical condition. Some women hear the grim news that their babies may die before birth. This occurs all too commonly: birth defects annually complicate an estimated 3% of all U.S. pregnancies and contribute to 20% of all infant deaths.[1] After receiving such a diagnosis, the pregnant woman typically receives a tremendous amount of medical information regarding her two possible choices: continuing her pregnancy with ongoing expectant management or pregnancy termination. Though the overall incidence of serious birth defects has remained relatively stable over time, medical knowledge regarding prognosis and therapy for many serious congenital conditions has greatly expanded over the past decade. This has led to changes in the way professionals counsel and support families. Several serious congenital conditions, until recently regarded as uniformly fatal, now respond to treatments that can extend life for weeks, months or years. Reports of long-term survivors with so-called "lethal" conditions highlight the considerable ambiguity inherent in prenatal prognostication. In this discussion we focus on families dealing with this uncertainty amidst their grief. How do medical providers and families approach decision-making in perinatal palliative care, and how has this changed in recent years? How does increasing parent autonomy in a changing social environment better inform care for families struggling with adverse prenatal diagnoses?

Perinatal palliative care

Perinatal palliative care is a developing field that aims to provide multidisciplinary and comprehensive patient- and family-centered support for decision-making and care coordination after a concerning fetal diagnosis. Perinatal palliative care providers seek to help expectant parents and families digest overwhelming information so that they can better formulate and enact personalized plans of care.[2] Focused palliative care in the perinatal period originated relatively recently and applied principles of modern adult

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hospice care to the prenatal period and beyond. Hoeldtke et al.[3] described their work caring for women who received fetal diagnoses predicted as lethal and chose to continue their pregnancies. The authors noted that obstetrical care teams often strongly recommended pregnancy termination, placing women who chose expectant management in a professional blind spot. Subsequently, a number of different reports described implementation of formal perinatal hospice programs, centered on providing postpartum comfort care to newborns after diagnoses with well-established high rates of intrauterine fetal demise or early postnatal death.[4–7]

Ongoing advances in prenatal screening and diagnosis have brought increasing complexity to prenatal prognostication. Studies such as maternal–fetal MRI, cell-free fetal DNA testing of maternal blood, and advanced fetal genetic and genomic analyses have resulted in earlier and more frequent diagnoses of congenital anomalies prior to birth.[8] Multiple studies of communication during the perinatal period have shown that families want and benefit from definitive information delivered in a clear and empathic manner.[9,10] However, vague predictions may be less useful and may indeed cause secondary trauma after the initial diagnosis.[11] Interpretation of relatively new test results may prove challenging, as rapidly advancing diagnostic technologies may outpace the clinical understanding of patient outcomes. Medical and surgical interventions continue to grow in number and intensity, allowing improved survival and decreased morbidity for some infants. Without complete data on outcomes of infants with rare prenatally diagnosed conditions, clinicians cannot provide accurate prognostic and therapeutic counseling for parents. To illustrate how advancing medical knowledge and shifting practice may affect prenatal prognostication and family experiences, we examine the situation regarding children with trisomies 13 and 18.

Trisomies 13 and 18 and the changing landscape of perinatal palliative care

Parental experience of children with trisomies 13 and 18 may serve as a model for how social change and emerging evidence have altered perinatal palliative care, mirroring experiences with trisomy 21 in the late 20th century.[12,13] Since initial descriptions in the 1960s, trisomies 13 and 18 were commonly regarded as lethal conditions due to high pre- and postnatal mortality.[14,15] Population-based surveys of infants during this period reported median survival of live-born infants between a few days to weeks and survival beyond 1 year averaging 5–10%, accompanied by profound neurodevelop-mental disability.[16–19] Along with high rates of intrauterine and intrapartum demise,[20,21] these statistics prompted care for prenatally diagnosed cases to center on either pregnancy termination or maternally-focused obstetric management and nearly exclusive provision of comfort care for neonates.[16] In spite of this, case reports surfaced documenting long-term survival for both conditions.[17,19,22] A relatively thin evidence base regarding the factors that contribute to longevity complicated prognostication, with outcomes remaining poorly studied. As a result, clinical management proceeded based solely upon abnormal karyotypes.

Over the past decade, however, several intersecting medical and societal factors began to influence this practice. First, progressive advancement of neonatal intensive care prolonged the lives of many medically complex, fragile newborns. Several small case series from the mid-2000s, primarily from Japan, began to report the experiences of infants with trisomy 13 and 18 receiving neonatal intensive care and cardiac surgery. One center described a cohort of 24 infants with trisomy 18: 88% received mechanical ventilation, with a median survival of 152.5 days and survival rates of 83% at 1 month and 25% at 1 year. Congenital cardiac disease was the most common factor in mortality.[23] An analysis of intensive treatment for 16 neonates with trisomy 13 revealed median survival of 733 days with a bimodal pattern of 1-year survival: approximately half survived less than 2 months and the remainder lived for at least 1 year.[24] Subsequent series reported the impact of cardiac surgery on infants with both trisomy 13 and 18 and suggested improved overall survival with either palliative surgery or cardiac repair for of a variety of congenital heart defects.[25–27] Together these studies introduced the idea that, at least for some neonates, early interventions can substantially prolong survival. However, neonatal intensive care and surgery remained controversial in much of the world. Perinatal palliative care for these children continued to focus on comfort with hospice-based care, citing the "best interest of the child."[28]

At about the same time, the disability rights movement prompted changes in societal attitudes regarding the quality of life of those with severe disabilities, encouraging an emphasis on patient capacity and autonomy or, where appropriate, surrogate decision-making.[29,30] The rise of family advocacy organizations, accompanied by online support and social media, provided widespread access to a more positive outlook on life with surviving children. Surveys of parents of children with trisomies 13 or 18 who belong to support groups almost universally describe their child as happy, interactive with family members, and enriching family life.[30,31] Psychomotor assessments show that these children, while having severely disabilities, can achieve some developmental milestones.[32] Following initial diagnosis, families searching online encounter images of these children which counter providers' dire predictions of "certain" mortality or rare survivors in a "vegetative" state. Many parents who seek and find such online support feel empowered to advocate for more intensive medical care for their children. In this changing medical and social landscape, larger and more recent studies from North America have shown that children with both trisomies increasingly experience hospitalization, often receiving intensive medical and surgical interventions that may contribute to longer survival. One large study surveyed a U.S. database of in-hospital care for children with either trisomy 13 or 18.[33] One-third of hospitalizations represented birth admissions, with 46% discharged home alive. Another third of hospitalizations were for children over 1 year of age. Over 2500 major therapeutic interventions were performed over the 5 years of the study. A separate study from Canada indicated that more children with these conditions receive surgical care, ranging from minor procedures to major cardiac surgery.[34] While early mortality remained very common, survival rates appeared to stabilize for both