Values Beneath the Surface: Ethical Considerations in Interpreting Clinical Data for Children with Trisomy 13 and 18 Requiring Cardiac Surgery and Respiratory Support

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Abstract

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In the last decade, there has been a notable shift in the care of children with trisomy 13 (T13) and trisomy 18 (T18) that challenges the predominant view of nonintervention due to high early mortality rates in these cohorts. Recent literature suggests that the increased frequency of surgical interventions, increased parental involvement in decision-making, disability advocacy, family support groups, and social networks have influenced this paradigm shift. Given these factors, more children with T13 and T18 are receiving life-sustaining interventions and living longer. However, the heterogeneity of these genetic conditions and limitations of retrospective studies makes it difficult to determine the extent to which certain factors cause early mortality. Common factors associated with early mortality in patients with T13 and T18 are lung and airway diseases, abnormal respiratory control, and structural cardiac conditions. When congenital cardiac malformations and pulmonary disorders coincide, these pulmonary conditions

can increase the risk of morbidity and mortality following cardiac surgery. Respiratory support is frequently mentioned as a negative predictor of postoperative survival, but few studies focus on the effect of respiratory support on survival outcomes. The goals of this analysis were to (1) characterize current morbidity and mortality in pediatric patients with T13 and T18 undergoing cardiac surgery, focusing on ventilation as a risk factor for survival, and (2) explore ethical considerations surrounding cardiac surgery through a role-based lens. This analysis considers how survival data and their interpretations influence judgments towards providing interventions to children with T13 and T18 who are dependent on respiratory technology. The findings from this analysis indicate that a majority of children who received preoperative MV or postoperative tracheostomy survived to hospital discharge. From examining the clinical manuscripts, quality-of-life assessments and perceptions of dis/ability, futility, and successful outcomes are compared among the papers' conclusions. This analysis emphasizes how one's values and relationship with the child influence clinical decision-making, underscoring the need for shared decision-making among parents and clinicians with regard to cardiac surgery in children with T13 and T18 receiving respiratory support.

Acknowledgements

"Nothing that we do that is worthwhile is done alone." —Mariame Kaba

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To the numerous faculty and ELSI scholars who have mentored me, your commitment to my personal and academic growth has been invaluable. Thank you for inspiring me.

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To my family and friends, I would not be who I am or where I am today without your love, encouragement, and patience.

It has been an honor to contribute to this body of knowledge as a researcher with a non-visible disability, writing about a group with whom I have no direct experience with. My understanding has been shaped by my experiences, mentorship, and reading. I want to acknowledge that individuals with T13 and T18 and their families have been pivotal in leading this work, ensuring their voices are heard in shaping the direction and outcomes of research. Their passionate voices and advocacy make this research possible.

Preface

Pictures deliver a stronger message than words could convey and during my research, I sat with an image in Guon et al. 2013 (Figure 3).[1] It is a collage of children with T13 and T18 and their respective ages. Parents that participated in the study agreed to have images and names of their children published. The authors (my thesis chairperson included) presented a more representative spectrum of children with T13 and T18 than previously published in the medical literature. They reminded readers that children with T13 and T18 create meaningful relationships with family members, have happy childhoods, and reach milestones.

Around the same time I read the Guon et al. study, I was also reading *No Pity* by Joseph Shapiro. I was fortunate to take a disability law and policy course while writing my thesis, where I was exposed to new voices, like Shapiro's, and considerations in disability studies. Shapiro wrote about disabling images, ones that "are internalized by disabled and nondisabled people alike and *build social stereotypes, create artificial limitations, and contribute to the discrimination and minority status hated by most disabled people*."(emphasis added)[2]

Working closely with Dr. Wilfond and drawing inspiration from his passion for this subject, I have come to admire the profound influence of language and imagery in shaping perceptions about disability. Dr. Wilfond spoke about the power of these elements to disrupt the stereotypes that label children with certain genetic diagnoses as "incompatible with life." This thesis serves as a greater appreciation of the inherent worth and potential within each of us as we challenge society's traditional notions of "health" and redefine what it truly means to be valued as human beings.

[1] Jennifer Guon et al., "Our Children Are Not a Diagnosis: The Experience of Parents Who Continue Their Pregnancy after a Prenatal Diagnosis of Trisomy 13 or 18," American Journal of Medical Genetics Part A 164, no. 2 (February 2014): 308–18, https://doi.org/10.1002/ajmg.a.36298.
[2] Shapiro, Joseph P. No Pity: People with Disabilities Forging a New Civil Rights Movement. (New York: Times Books, 1993), 30.

Table of Contents

1. Introduction

1.1. History and significance of trisomy 13 and 18

Trisomy 13 (T13) and trisomy 18 (T18) are the most common autosomal chromosomal disorders in newborns after trisomy 21, with an estimated prevalence of live borns with T13 and T18 of 1 in 10,000 and 1 in 6,000, respectively.[1] The total birth prevalence[2] (live births, stillbirths, and terminations) is about 1 in 6,200 total births for T13 and about 1 in 2,500 total births for T18.[3] A range of congenital anomalies characterize T13 and T18, including multiorgan malformations, central nervous system defects, and cardiopulmonary disorders.[4] Cardiac malformations are the most frequent anomalies seen in children with T13 and T18, with congenital heart disease presenting in 80-90% of patients and ventricular septal defects (VSD) as the primary cardiac defect.[5]

The combination of morbidities in T13 and T18 are associated with poor prognosis and high infant mortality, criteria that have supported the labeling of these conditions as "lethal" anomalies.[6] The median age of survival for children with T13 and T18 is between 1–3 weeks, although survival rates are increasing for children who receive medical interventions.[7] Studies report many children with T13 and T18 being discharged from the hospital and a small

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[1] John C. Carey, "TRISOMY 18 AND TRISOMY 13 SYNDROMES," in *Cassidy and Allanson's Management of Genetic Syndromes*, ed. John C. Carey et al., 4th ed. (Wiley, 2021), 937–56, https://doi.org/10.1002/9781119432692.ch58.

[2] The true incidence of these conditions from conception onward is unknown because it is difficult to ascertain complete and accurate data on fetal deaths and pregnancy terminations due to T13 and T18. For example, not all pregnant individuals receive prenatal diagnoses, and genetic testing for chromosomal anomalies after a miscarriage is not routinely performed.

[3] Carey, "TRISOMY 18 AND TRISOMY 13 SYNDROMES."

[4] Ibid.

[5] Ibid.

[6] Tracy K. Koogler, Benjamin S. Wilfond, and Lainie Friedman Ross, "Lethal Language, Lethal Decisions," *The Hastings Center Report* 33, no. 2 (March 2003): 37, https://doi.org/10.2307/3528153.

[7] John C. Carey, "Survival Outcomes of Infants with the Trisomy 13 or Trisomy 18 Syndromes," *The Journal of Pediatrics* 247 (August 1, 2022): 11–13, https://doi.org/10.1016/j.jpeds.2022.05.043; Katherine A. Kosiv et al., "A Validated Model for Prediction of Survival to 6 Months in Patients with Trisomy 13 and 18," *American Journal of Medical Genetics Part A* 185, no. 3 (2021): 806–13, https://doi.org/10.1002/ajmg.a.62044.

percentage living well into childhood.[8] Despite high in-hospital mortality, one long-term study reported that 10-year survival could be achievable in patients with T13 and T18 following cardiac surgery.[9] A recent European cohort study also confirms longer survival than previously reported for children with T13 and T18, finding that 10-16% of children survived to 5 years of age and 8-11% survived to 10 years of age.[10]

Historically, policy guidelines from professional medical organizations indicated that resuscitation and other life-sustaining interventions were not indicated for so-called lethal anomalies such as T13 and T18.[11] High early mortality and concerns about quality of life justified widespread nonintervention policies, presumably in the best interest of the child given the probability of reduced neurocognitive ability.[12] This became a self-fulfilling prophecy, where the rarity of providing life-sustaining interventions reinforced low survival rates.[13] Many scholars have called attention to how values shape and sometimes conflate treatment recommendations and "turn normative judgments into clinical ones."[14]

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[8] Horacio G. Carvajal et al., "Cardiac Surgery in Trisomy 13 and 18: A Guide to Clinical Decision-Making," *Pediatric Cardiology* 41, no. 7 (October 2020): 1319–33, https://doi.org/10.1007/s00246-020-02444-6.
[9] Jennifer K. Peterson et al., "Long Term Outcomes of Children with Trisomy 13 and 18 After Congenital Heart Disease Interventions," *The Annals of Thoracic Surgery* 103, no. 6 (June 2017): 1941–49, https://doi.org/10.1016/j.athoracsur.2017.02.068.
[10] Svetlana V. Glinianaia et al., "Ten-Year Survival of Children with Trisomy 13 or Trisomy 18: A Multi-Registry European Cohort Study," *Archives of Disease in Childhood*, March 7, 2023, https://doi.org/10.1136/archdischild-2022-325068.
[11] Koogler, Wilfond, and Ross, "Lethal Language, Lethal Decisions"; Jennifer Guon et al., "Our Children Are Not a Diagnosis: The Experience of Parents Who Continue Their Pregnancy after a Prenatal Diagnosis of Trisomy 13 or 18," *American Journal of Medical Genetics Part A* 164, no. 2 (February 2014): 308–18, https://doi.org/10.1002/ajmg.a.36298.
[12] Koogler, Wilfond, and Ross, "Lethal Language, Lethal Decisions"; Melanie P. McGraw and Jeffrey M. Perlman, "Attitudes of Neonatologists Toward Delivery Room Management of Confirmed Trisomy 18: Potential Factors Influencing a Changing Dynamic," *Pediatrics* 121, no. 6 (June 1, 2008): 1106–10, https://doi.org/10.1542/peds.2007-1869.
[13] John P. Cleary et al., "Cardiac Interventions for Patients With Trisomy 13 and Trisomy 18: Experience, Ethical Issues, Communication, and the Case for Individualized Family-Centered Care," *World Journal for Pediatric and Congenital Heart Surgery* 13, no. 1 (January 1, 2022): 72–76, https://doi.org/10.1177/21501351211044132.
[14] Koogler, Wilfond, and Ross, "Lethal Language, Lethal Decisions"; Annie Janvier, Barbara Farlow, and Keith Barrington, "Cardiac Surgery for Children with Trisomies 13 and 18: Where Are We Now?," *Seminars in Perinatology* 40, no. 4 (June 2016): 254–60, https://doi.org/10.1053/j.semperi.2015.12.015; Annie Janvier and Andrew Watkins, "Medical Interventions for Children with Trisomy 13 and Trisomy 18: What Is the Value of a Short Disabled Life?," *Acta Paediatrica* 102, no. 12 (December 2013): 1112–17, https://doi.org/10.1111/apa.12424;

This standard of nonintervention in children who have T13 and T18 parallels the treatment of Babies Jane and John Doe in the 1980s.[15] At the advice of the respective medical teams, both parents declined surgical interventions but the cases had vastly different outcomes: Baby Jane Doe, born with spina bifida, passed her 30th birthday in 2013, but Baby John Doe, born with trisomy 21, died six days after birth.[16] Fierce ethical and legal debates surrounding the Baby Doe cases led to the enactment of federal protections for vulnerable children and underscored the polarity between medicine and disability rights. The medical and genetic model regards disability as an individual condition or difference that can be fixed or prevented.[17] There is an ideal state of health, an illusion of a "normal," functioning body, to be achieved and managed with medicine. The medicalization and stigmatization of disability have roots in the eugenics movement dating back to the late 19th century. Eugenicists aimed to reduce and marginalize populations deemed inferior and unfit, including eliminating disability from the population.[18]

The social or civil rights model regards disability as a sociopolitical and economic construct, and it is society, not the individual, that should change.[19] The social model, unlike the medical model, is less concerned with pathology; rather, disability is circumstantial and seen as

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Guon et al., "Our Children Are Not a Diagnosis"; Benjamin S. Wilfond and John C. Carey, "Parental Requests for Intervention in Children with Lethal Conditions," in *Clinical Ethics in Pediatrics*, ed. Douglas S. Diekema, Mark R. Mercurio, and Mary B. Adam, 1st ed. (Cambridge University Press, 2011), 174–80, https://doi.org/10.1017/CBO9780511740336.031.

[15] Alicia Ouellette, *Bioethics and Disability: Toward a Disability-Conscious Bioethics* (Cambridge: Cambridge University Press, 2011), 23.

[16] Ibid.; Adrienne Asch, "Disability, Bioethics and Human Rights," in *Handbook of Disability Studies* (Thousand Oaks, Calif: Sage Publications, 2001); Nicole Fuller, "'Baby Jane Doe' at 30: Happy, Joking, Learning - Newsday," Newsday, October 13, 2013, https://www.newsday.com/news/health/baby-jane-doe-at-30-happy-joking-learning-u93680.

[17] Ouellette, *Bioethics and Disability*, 58; Jackie Leach Scully, *Disability Bioethics: Moral Bodies, Moral Difference*, Feminist Constructions (Lanham: Rowman & Littlefield, 2008), 22–25.

[18] Ibid.

[19] Ibid.; Paul Steven Miller and Rebecca Leah Levine, "Avoiding Genetic Genocide: Understanding Good Intentions and Eugenics in the Complex Dialogue between the Medical and Disability Communities," *Genetics in Medicine* 15, no. 2 (February 2013): 95–102, https://doi.org/10.1038/gim.2012.102.

the result of a dysfunctional society. Instead, "people with disabilities are demanding rights, not medical cures."[20] Importantly, these models are generalizations of different conceptions of disability, each with its own variations and flaws; neither model can nor should be assumed to fully explain a person's views or actions.

Justification for withholding interventions from children with T13 and T18 relies on the medical model of disability: life-limiting genetic conditions are not curable and thus treatment could be considered "futile." Nonintervention can be permissible to avoid prolonging an inevitable death or harming a child with intensive care that might result in them having a technology-dependent life.[21] But death does not have to be inevitable for children with T13 and T18. Each child is unique with respect to their genetics, predisposing factors, and the goals and values of their parents and clinicians.[22]

A notable shift in frequency and level of care for children with T13 and T18 has been described in the literature, including an increase in major cardiac surgeries as opposed to palliative or comfort care.[23] This reflects clinicians being more receptive to providing interventions in these cohorts, including high-risk children receiving artificial ventilation.[24] Scholars suggest that the increased frequency of surgical interventions, increased parental

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[20] Shapiro, Joseph P. *No Pity: People with Disabilities Forging a New Civil Rights Movement.* (New York: Times Books, 1993), 14.
[21] Chris Feudtner et al., "Technology-Dependency among Patients Discharged from a Children's Hospital: A Retrospective Cohort Study," *BMC Pediatrics* 5, no. 1 (May 9, 2005): 8, https://doi.org/10.1186/1471-2431-5-8; Janvier and Watkins, "Medical Interventions for Children with Trisomy 13 and Trisomy 18."
[22] Guon et al., "Our Children Are Not a Diagnosis."
[23] John C. Carey, "Management of Children with the Trisomy 18 and Trisomy 13 Syndromes: Is There a Shift in the Paradigm of Care?," *American Journal of Perinatology* 38, no. 11 (September 2021): 1122–25, https://doi.org/10.1055/s-0041-1732363; Katherine E. Nelson, Kari R. Hexem, and Chris Feudtner, "Inpatient Hospital Care of Children With Trisomy 13 and Trisomy 18 in the United States," *Pediatrics* 129, no. 5 (May 1, 2012): 869–76, https://doi.org/10.1542/peds.2011-2139; Meaghann S. Weaver et al., "Mixed Method Study of Quality of Life for Children with Trisomy 18 and 13 after Cardiac Surgery," *Cardiology in the Young* 30, no. 2 (February 2020): 231–37, https://doi.org/10.1017/S1047951120000013.
[24] Carey, "TRISOMY 18 AND TRISOMY 13 SYNDROMES"; Joshua M. Rosenblum et al., "Cardiac Surgery in Children with Trisomy 13 or Trisomy 18: How Safe Is It?," *JTCVS Open*, September 2022, S2666273622003564, https://doi.org/10.1016/j.xjon.2022.09.005.

involvement in decision-making, disability advocacy, family support groups, and social networks have contributed to increased surgical operations and survival rates.[25] This paradigm shift challenges the predominant view of nonintervention, though the evolution of practice is not uniform; hospitals may shy away from providing treatment in complex cases where patients require respiratory support because mechanical ventilation (MV) has been widely reported as a negative predictor of survival over the last two decades.

1.2. Study purpose

"It is often said in clinical ethics that good ethics starts with good facts. With [T13 and T18], we can observe that values influence medical statistics."[26]

Respiratory anomalies and complications, such as MV, tracheostomy, and infection, are frequently seen in children with T13 and T18.[27] Cooper et al. provided evidence that MV is associated with increased postoperative mortality and is therefore a contraindication to cardiac surgery.[28] Further, the inability to extubate children and the small number of intubated children who are discharged home supports futility arguments—that aggressive, life-sustaining interventions are not beneficial in the long term and are morally wrong.[29] Respiratory conditions are a major cause of death in children with T13 and T18 and airway management is critical to

[25] Carey, "Management of Children with the Trisomy 18 and Trisomy 13 Syndromes"; Annie Janvier, Barbara Farlow, and Benjamin S. Wilfond, "The Experience of Families With Children With Trisomy 13 and 18 in Social Networks," *Pediatrics* 130, no. 2 (August 1, 2012): 293–98, https://doi.org/10.1542/peds.2012-0151.
[26] Janvier and Watkins, "Medical Interventions for Children with Trisomy 13 and Trisomy 18."
[27] Carey, "TRISOMY 18 AND TRISOMY 13 SYNDROMES"; Sara K. Swanson et al., "Impact of Trisomy 13 and 18 on Airway Anomalies and Pulmonary Complications after Cardiac Surgery," *The Journal of Thoracic and Cardiovascular Surgery* 162, no. 1 (July 2021): 241–49, https://doi.org/10.1016/j.jtcvs.2020.08.082.
[28] David S. Cooper et al., "Cardiac Surgery in Patients With Trisomy 13 and 18: An Analysis of The Society of Thoracic Surgeons Congenital Heart Surgery Database," *Journal of the American Heart Association: Cardiovascular and Cerebrovascular Disease* 8, no. 13 (June 25, 2019): e012349, https://doi.org/10.1161/JAHA.119.012349.
[29] Eric M. Graham, "Infants with Trisomy 18 and Complex Congenital Heart Defects Should Not Undergo Open Heart Surgery," *Journal of Law, Medicine & Ethics* 44, no. 2 (2016): 286–91, https://doi.org/10.1177/1073110516654122.

their care.[30] Yet respiratory support is not necessarily against a child's best interests.[31] In fact, MV has been predicted to be protective against cardiac surgery-associated mortality.[32]

The role of respiratory support is an important consideration in the decision-making process between two relevant interested parties: parents of children with T13 and T18 and the clinicians that assume their care. However, parents and clinicians interpret clinical outcomes differently based on deeply held values and beliefs on disability, meaningful medical interventions, and the best interests of the child.[33] As one article pointed out, higher survival rates for children with T13 and T18 are more likely attributed to differing values than differing medical approaches or resources.[34]

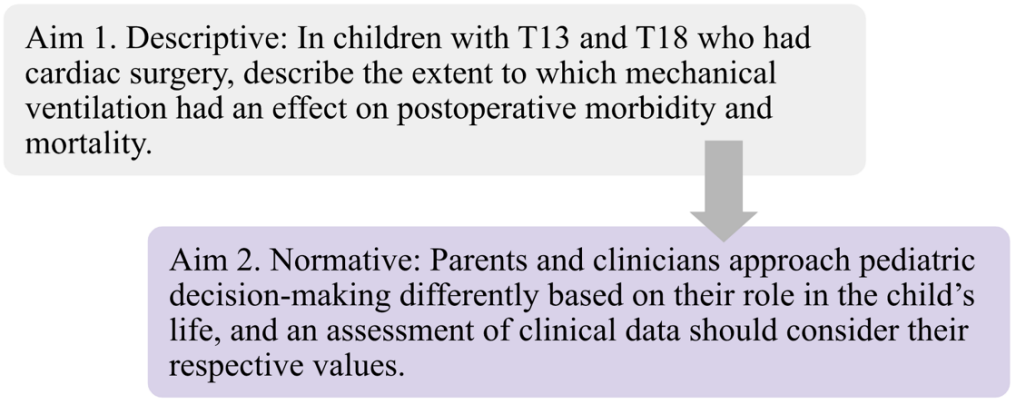
The objective of this analysis was to evaluate current epidemiological data of children with T13 and T18 who received perioperative assisted ventilation and cardiac surgery to support further consideration of how these data and their interpretations might influence clinical decision-making. When subjective terminology, such as incompatibility with life, lethality, and quality of life, is used in conjunction with clinical data, cardiac surgery appears to be counterintuitive and futile in these cohorts. But children with T13 and T18 receiving respiratory support can live a meaningful life because "meaning" and "worth" are specific to each family. It is not obligatory that all children with T13 and T18 receive cardiac surgery; however, this thesis concludes that it is unjust to deny surgery based solely on their respiratory support status.

[30] Carey, "TRISOMY 18 AND TRISOMY 13 SYNDROMES."
[31] Benjamin S. Wilfond, "Tracheostomies and Assisted Ventilation in Children With Profound Disabilities: Navigating Family and Professional Values," *Pediatrics* 133, no. Supplement_1 (February 1, 2014): S44–49, https://doi.org/10.1542/peds.2013-3608H.
[32] Kosiv et al., "A Validated Model for Prediction of Survival to 6 Months in Patients with Trisomy 13 and 18."
[33] Wilfond, "Tracheostomies and Assisted Ventilation in Children With Profound Disabilities"; Janvier, Farlow, and Wilfond, "The Experience of Families With Children With Trisomy 13 and 18 in Social Networks."
[34] Janvier, Farlow, and Barrington, "Cardiac Surgery for Children with Trisomies 13 and 18."

1.3. Specific aims

The specific aims of this analysis were to (1) characterize the data regarding current morbidity and mortality in pediatric patients with T13 and T18 who underwent cardiac surgery and received perioperative respiratory support, and (2) explore ethical considerations related to cardiac surgery decisions for patients dependent on respiratory support from a role-differentiated perspective (Figure 1).

Figure 1. The relationship between aims



2. Approach

A review of contemporary data describing perioperative respiratory support is necessary to contextualize an analysis of ethical considerations in these cohorts. Evidence and conclusions from the literature guide clinical decision-making, a process that relies on values as much as data.[35] Commenting on data without addressing the implicit or explicit values that shape them

[35] Mark R. Mercurio, "The Ethics of Newborn Resuscitation," *Seminars in Perinatology* 33, no. 6 (December 2009): 354–63, https://doi.org/10.1053/j.semperi.2009.07.002.

would be unremarkable.[36] Therefore, the approach for this analysis was two-fold, as described below.

2.1. Mixed purposeful sampling of epidemiological studies

Given the clinical nature and specificity of the research objective, the PICo model (Population, Interest, Context) was used to guide the research question and search strategy, from which criterion and emergent sampling could be leveraged to evaluate the resulting articles.[37] This analysis was interested in morbidity and mortality (Interest) in children with T13 and T18 with heart anomalies who underwent cardiac surgery in the United States (Population) and received perioperative respiratory support (specifically high flow nasal cannula (HFNC), MV, or tracheostomy) (Context).

Purposeful sampling is a method of qualitative research synthesis that focuses on information-rich sources.[38] There are several strategies in purposeful sampling, two of which were used in this study: criterion sampling and emergent sampling. Criterion sampling identifies cases from preset criteria, while emergent sampling incorporates cases ad hoc.[39]

The PubMed and Web of Science databases were queried using the following search string, "(Trisomy 13 AND Trisomy 18) AND cardiac surgery AND (mechanical ventilation OR respiratory OR respiration)", with a date range of 2000-2023; this returned 14 distinct articles.[40]

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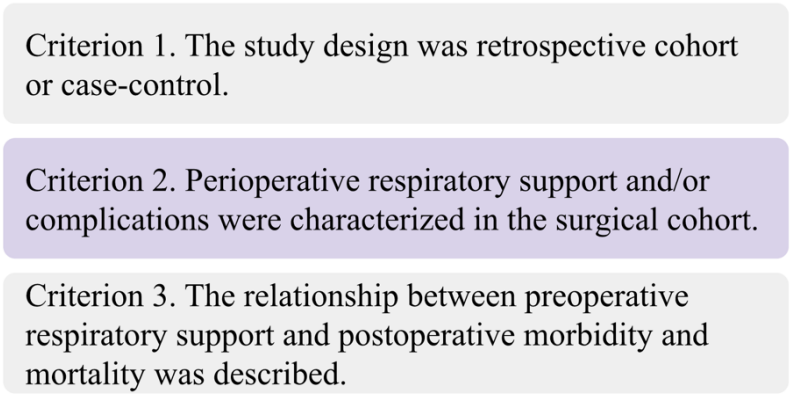
[36] Gregory E. Kaebnick, "Civic Learning When the Facts Are Politicized: *How Values Shape Facts, and What to Do about It*," *Hastings Center Report* 51, no. S1 (January 2021), https://doi.org/10.1002/hast.1228.
[37] Cindy Stern, Zoe Jordan, and Alexa McArthur, "Developing the Review Question and Inclusion Criteria," *AJN The American Journal of Nursing* 114, no. 4 (April 2014): 53, https://doi.org/10.1097/01.NAJ.0000445689.67800.86.
[38] Harsh Suri, "Purposeful Sampling in Qualitative Research Synthesis," *Qualitative Research Journal* 11, no. 2 (August 3, 2011): 63–75, https://doi.org/10.3316/QRJ1102063.
[39] Ibid.
[40] PubMed was queried first (N=8), then Web of Science (N=13). Duplicates from Web of Science were excluded (N=7).

From this set, articles were reviewed and included if they met three predetermined criteria (Figure 2).

Figure 2. Criterion sampling for studies pertaining to children with T13 and T18 who underwent cardiac surgery and received preoperative or postoperative respiratory support



Application of these criteria yielded four articles. Review articles, duplicate articles, and quantitative studies unrelated to the population or intervention were excluded. However, one review article[41] found in both databases summarized studies that characterized T13 and T18 cardiac surgery cohorts, outcomes, and perioperative risk factors between 2004-2020. While not an epidemiological study, this emergent article was narrowly tailored to the research objective and presented 13 additional epidemiological studies not identified through the initial criterion sampling. In sum, 27 articles were screened according to the criteria in Figure 2 (PubMed and Web of Science=14; Carvajal et al. 2020=13) and five articles met the inclusion criteria (PubMed=3; Web of Science=1; Carvajal et al.=1).

[41] Carvajal et al., "Cardiac Surgery in Trisomy 13 and 18."

2.2. Roles and ethical considerations in pediatric decision-making

"These are the kinds of clinical ethics cases where clinicians ask, "How can the parents do this to their child?" and parents ask, "How can we do anything else?"[42]

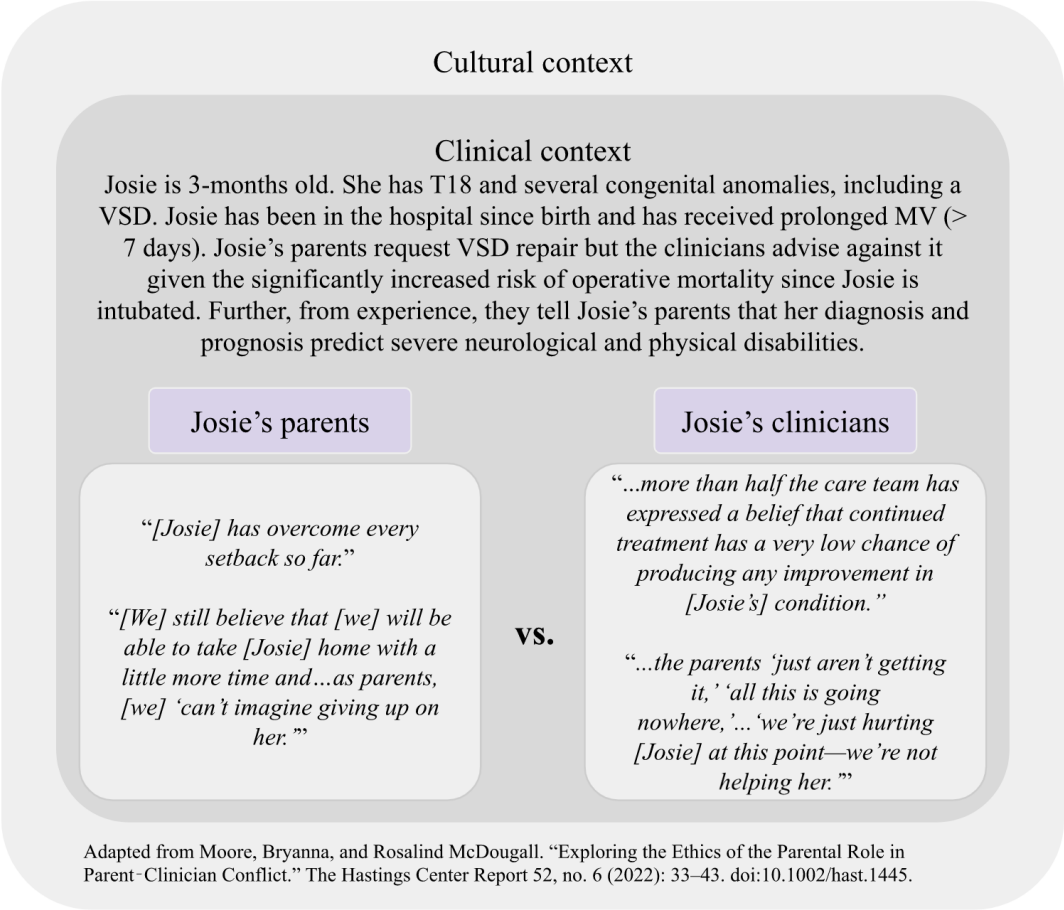
A number of pediatric ethics frameworks can be used to guide clinical decision-making in children with disabilities; this analysis focused on a role-differentiated lens of parents and clinicians caring for children with T13 and T18. A role-differentiated approach aims to clarify why parents and clinicians value decisions and clinical outcomes differently while sharing similar goals. The roles of parents and clinicians are distinct and shaped by an understanding of what their "right action" is given their relationship with the child.[43] Values and roles are critical components to a shared decision-making (SDM) approach; this model, a collaborative, relationship-focused process to care management, has been described in the context of children with T13 and T18 throughout different decision points in prenatal and postnatal care.[44]

In addition to the legal and social weight parents carry in pediatric decision-making, a role-based approach attempts to hold the parental role at an equitable ethical weight to the clinician role (Figure 3).[45] This is important because parents, while adapting to a potentially unfamiliar situation of caring for a critically ill child, are also trying to maintain what they believe are the roles of a "good parent."[46] SDM is one strategy to elevate and maintain equitable parental weight when ethical tensions arise in the determination of what medical interventions, if

[42] Bryanna Moore and Rosalind McDougall, "Exploring the Ethics of the Parental Role in Parent-Clinician Conflict," *Hastings Center Report* 52, no. 6 (November 2022): 33–43, https://doi.org/10.1002/hast.1445.
[43] Ibid.
[44] Sasha E. Andrews et al., "Shared Decision Making and the Pathways Approach in the Prenatal and Postnatal Management of the Trisomy 13 and Trisomy 18 Syndromes," *American Journal of Medical Genetics Part C: Seminars in Medical Genetics* 172, no. 3 (2016): 257–63, https://doi.org/10.1002/ajmg.c.31524.SD
[45] Moore and McDougall, "Exploring the Ethics of the Parental Role in Parent-Clinician Conflict."
[46] Ibid.

any, are in the child's best interests.[47] What parents see as a commitment to their child (e.g., advocating for life-sustaining treatment; a good parent never "gives up"), clinicians might see as a burden to provide the best care for their patient (e.g., preventing suffering or harm to the child; parents do not understand the complexity of these situations). The authors of the role-based framework note that restructuring conflict to appreciate why parents and clinicians come to different conclusions can promote respectful engagement during distressing situations.[48]

Figure 3. Parent and clinician perspectives in cardiac surgery candidacy for patients with T13 and T18 receiving respiratory support



Adapted from Moore, Bryanna, and Rosalind McDougall. "Exploring the Ethics of the Parental Role in Parent‐Clinician Conflict." The Hastings Center Report 52, no. 6 (2022): 33–43. doi:10.1002/hast.1445.

[47] Andrews et al., "Shared Decision Making and the Pathways Approach in the Prenatal and Postnatal Management of the Trisomy 13 and Trisomy 18 Syndromes."
[48] Moore and McDougall, "Exploring the Ethics of the Parental Role in Parent-Clinician Conflict."

An evaluation of epidemiological data relies on an understanding of ethical considerations for children with T13 and T18 who are mechanically ventilated and potential candidates for cardiac surgery. Parents and clinicians contemplate the same ethical considerations (such as survival, quality of life, and suffering) based on their distinct values and responsibilities to the child.[49] The considerations discussed in this analysis (Table 1) were adapted from ethical issues in the context of clinical decision-making for children with perceived lethal anomalies.[50] The epidemiological articles previously identified and any corresponding commentaries[51] were reviewed in light of these considerations.

Table 1. Ethical considerations for parents and clinicians regarding cardiac surgery in children with T13 and T18 with congenital heart anomalies and respiratory support

Considerations	**Description**
Quality of life	Value judgments assigned to an assessment of benefits and burdens for an intervention in a determination of what is acceptable and unacceptable
Attitudes about dis/ability[52]	Presumptions of the child's ability and disability
Quantitative futility	Justification for withholding an intervention based on a low probability of it being successful
Successful surgical outcomes	Value assigned to postoperative outcomes such as hospital discharge and survival

[49] Kathryn Neubauer and Renee D. Boss, "Ethical Considerations for Cardiac Surgical Interventions in Children with Trisomy 13 and Trisomy 18," *American Journal of Medical Genetics Part C: Seminars in Medical Genetics* 184, no. 1 (2020): 187–91, https://doi.org/10.1002/ajmg.c.31767.
[50] Wilfond and Carey, "Parental Requests for Intervention in Children with Lethal Conditions."
[51] Included commentaries (N=2) were those listed under the parent PMID (N=5) in PubMed.
[52] The word "disability" has several constructions. Dis/ability surfaced as a new theory in Dan Goodley's 2014 book, *Dis/ability Studies: Theorising Disablism and Ableism*, to emphasize the coexistence of ability and disability as produced in the evolution of global politics; one cannot exist without the other. Other constructions include (dis)ability (See Sami Schalk, 2018, *Bodyminds Reimagined: (Dis)ability, Race and Gender in Black Women's Speculative Fiction*) and ability/disability (See Rosemarie Garland-Thomson, 2002, "Integrating Disability, Transforming Feminist Theory") to underscore the intersectionality between disability, race, gender, sexuality, age, class, and other social concepts.

3. Analysis

3.1. Epidemiological data regarding children with mechanical ventilation and cardiac surgery

A review of epidemiological data using purposeful sampling enabled the selection of studies that presented outcomes for children with T13 and T18 who received perioperative ventilation. Studies are summarized, with patient demographics and outcomes compared, in appendices A and B.

There were two multicenter retrospective cohort studies and three single-center retrospective studies, one case-control and two cohort. VSD repair was the most common cardiac procedure in each study. Of the two multicenter cohort studies, one reviewed cardiac operations over a 7-year period (Cooper et al. 2019) and the other reviewed operations over an 18-year period (Graham et al. 2004) (Appendix A).[53] Cooper et al. was the largest study to report epidemiological outcomes in cardiac surgery patients with T13 and T18 using the national Society of Thoracic Surgeons Congenital Heart Surgery Database. There were 73 operations on patients with T13 and 270 operations on patients with T18, with 16 and 82 of them receiving preoperative MV, respectively.

Graham et al. was a seminal study in 2004 that reported 35 operations on patients with T13 and T18 and tracked MV among two groups (0-2 days and >2 days) from before surgery through hospital discharge. Almost 75% of the cohort was ventilated for two days or fewer, with all surviving patients extubated before discharge. Conversely, more than half of the patients ventilated for more than two days were ventilated at discharge.

[53] Cooper et al., "Cardiac Surgery in Patients With Trisomy 13 and 18"; Eric M. Graham et al., "Effectiveness of Cardiac Surgery in Trisomies 13 and 18 (from the Pediatric Cardiac Care Consortium)," *The American Journal of Cardiology* 93, no. 6 (March 15, 2004): 801–3, https://doi.org/10.1016/j.amjcard.2003.12.012.

Among the single-center cohort studies, one reported nine patients over a 12-year period at their institution who had T18 and cardiac surgery (Davisson et al. 2018); a third of whom received preoperative MV and over half of whom had a postoperative tracheostomy.[54] The other single-center cohort study (Rosenblum et al. 2022) reported 19 operations over a 19-year period at their institution;[55] five patients never left the hospital, two received preoperative tracheostomies, and six received HFNC.[56] There was one single-center case-control study (Swanson et al. 2021) that described pre- and postoperative respiratory support in 14 patients who had cardiac surgery over a 20-year period.[57] Most of the patients at this institution had persistent preoperative hospitalization. In addition, five patients received preoperative respiratory support (four had MV and one had HFNC) and a majority had a form of postoperative respiratory support (seven had prolonged MV, two had tracheostomies, and two had HFNC).

Interestingly, a majority of patients who received preoperative MV survived to discharge, with the multicenter studies reporting survival between 63-92% and the single-center studies reporting survival between 67-75% (Appendix B). Of the two studies that reported postoperative tracheostomy placement, most patients survived to discharge (80-100%); however, these were both single-center studies with small cohorts of less than five patients with tracheostomies.

For both pre- and postoperative respiratory support, in-hospital mortality was categorized as within 30 days of surgery or before hospital discharge. One study did not report survival outcomes in patients who received preoperative respiratory support (Rosenblum et al.). Mortality associated with preoperative MV within 30 days of surgery was reported in one single-center

[54] Neena A. Davisson et al., "Trisomy 18 and Congenital Heart Disease: Single-Center Review of Outcomes and Parental Perspectives," *World Journal for Pediatric and Congenital Heart Surgery* 9, no. 5 (September 2018): 550–56, https://doi.org/10.1177/2150135118782145.
[55] Rosenblum et al., "Cardiac Surgery in Children with Trisomy 13 or Trisomy 18."
[56] For this analysis, HFNC is defined as greater than 1 liter per minute of supplemental oxygen.
[57] Swanson et al., "Impact of Trisomy 13 and 18 on Airway Anomalies and Pulmonary Complications after Cardiac Surgery."

study (Swanson et al., N=1, 25%) and one multicenter study (Graham et al., N=1 for patients with MV >2 days, 11%; N=2 for patients with MV 0-2 days, 8%). One study reported mortality associated with preoperative MV before hospital discharge (Davisson et al., N=1, 33%) and another did not differentiate in-hospital mortality (Cooper et al., N=5 for patients with T13, 31%; N=30 for patients with T18, 37%). Two single-center studies reported mortality associated with postoperative tracheostomy placement: Swanson et al. reported 0% mortality and Davisson et al. reported 20% mortality before discharge (N=1).

3.2. "The politics of probability": how values and roles shape the understanding of data

Four of the five epidemiological studies in this analysis described postoperative mortality in T13 and T18 cohorts, specifying which patients received perioperative respiratory support. In addition to reframing the approach through which parents and clinicians resolve conflicts in decision-making, reframing the data also tells an interesting story. While parents and clinicians share a commitment to care for and not harm the child, they will interpret clinical data differently.

Consider a scenario using data from Cooper et al. regarding children with T13 who underwent cardiac surgery: of the patients who survived their operations (N=65), 17% were mechanically ventilated before surgery (N=11) and of those who did not survive their operations (N=8), 63% were mechanically ventilated before surgery (N=5).[58] From these data, the authors concluded that preoperative MV was strongly associated with in-hospital mortality and therefore,

[58] Cooper et al., "Cardiac Surgery in Patients With Trisomy 13 and 18."

"this subset of patients with T13 and T18 should perhaps not be considered surgical candidates."[59]

However, these data also show that of the patients with T13 who received preoperative MV (N=16), 69% of the children survived (N=11). How might clinicians present this evidence to worrying parents in a way that conveys its importance? Are these data sufficient for clinicians, particularly cardiac surgeons, to reassess prioritizing parents' goals and wishes for their children? How do the values of parents and clinicians, shaped by their relationship with the child, factor into conversations of whether a child with T13 or T18 receiving artificial ventilation should also receive cardiac surgery?

When parents are told the odds are against their child surviving surgery due to their dependence on MV, it may be seldom mentioned that a clinician's assessment of mortality data also reflects their own values. Subjective factors such as personal beliefs and social roles shape one's understanding of objective facts, and this understanding has the potential to reinforce self-fulfilling prophecies.[60] "The politics of probability" acknowledges that statistics do not exist in a vacuum and contrasting interpretations of data will influence how they are used in clinical decision-making.[61]

This ethical analysis considers why parents and clinicians might come to different interpretations of ethical considerations and statistics associated with cardiac surgery in children with T13 and T18 who received respiratory support. The authors' conclusions from the selected epidemiological studies and commentaries are incorporated to show these contrasting

[59] Ibid.
[60] John D. Lantos, "Trisomy 13 and 18—Treatment Decisions in a Stable Gray Zone," *JAMA* 316, no. 4 (July 26, 2016): 396–98, https://doi.org/10.1001/jama.2016.9470.
[61] Joel Michael Reynolds, "May the Odds Be Ever in Your Favour? The Politics of Prognosis," *Aeon* (blog), March 5, 2018, https://aeon.co/ideas/may-the-odds-be-ever-in-your-favour-the-politics-of-prognosis.

perspectives, even among clinicians. While these papers did not include parental narratives, they provided valuable insight into how clinicians perceive the parent-child relationship. Ultimately, this analysis joins previous scholarship in advocating for clinicians to consider parents' values in pediatric SDM.[62]

3.2.1. Quality of life and attitudes about dis/ability

In a determination of whether surgical interventions should be performed in patients with T13 and T18, quality-of-life assessments typically surface during discussions about withholding treatment.[63] Some healthcare providers may hold a bias towards children with severe disabilities and not see their ability to interact and form meaningful relationships in the way parents and families do.[64] Parents have described negative interactions and differences of opinion with clinicians; oftentimes, there is an underlying presumption that the child's quality of life, even after the intervention, will be undesirable.[65] However, while survival statistics, medical assessments, and personal and professional values shape clinicians' recommendations, parents present a different perspective with different motivators.[66] Recognizing variations in decision-making by virtue of role is important to preserving the parent-clinician relationship and maximizing the potential of the child. While none of the five epidemiological studies explicitly mentioned "disability," statements regarding quality of life, as well as attitudes about the children's abilities, were found throughout the authors' conclusions.

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[62] Andrews et al., "Shared Decision Making and the Pathways Approach in the Prenatal and Postnatal Management of the Trisomy 13 and Trisomy 18 Syndromes."
[63] Wilfond and Carey, "Parental Requests for Intervention in Children with Lethal Conditions"; Janvier, Farlow, and Wilfond, "The Experience of Families With Children With Trisomy 13 and 18 in Social Networks."
[64] Guon et al., "Our Children Are Not a Diagnosis"; Janvier, Farlow, and Barrington, "Cardiac Surgery for Children with Trisomies 13 and 18."
[65] Janvier, Farlow, and Wilfond, "The Experience of Families With Children With Trisomy 13 and 18 in Social Networks"; Guon et al., "Our Children Are Not a Diagnosis."
[66] Janvier and Watkins, "Medical Interventions for Children with Trisomy 13 and Trisomy 18"; Moore and McDougall, "Exploring the Ethics of the Parental Role in Parent-Clinician Conflict."

The institutional policy discussed in Rosenblum et al. was to offer cardiac surgery to children with T13 and T18 under the condition that the child had been previously discharged.[67] They stated an initial preference to operate on children who demonstrated "a will to live [and] breathe." This choice of terminology is problematic as it assumes children with T13 and T18 who require MV do not express a wish or desire to breathe on their own. Value-driven language is not uncommon in healthcare. In this context, it introduces an unnecessarily restrictive criterion for surgical candidacy, where children with T13 and T18 may need to prove they are determined to live—an expectation seldom imposed on other pediatric patients. Similar to terms like "incompatible with life" and "vegetable," this terminology can be construed as underestimating and devaluing the ability of children with T13 and T18.

Further, parents have expressed their discontent with language that assumes their child's life has reduced value given a certain diagnosis and prognosis; parents appreciated healthcare providers using "child-first" language, or language that recognizes the uniqueness of their child.[68] The choice of language is powerful, particularly when discussing children with disabilities, as it can reflect one's beliefs about the quality of life for a child with different abilities, outcomes, and life expectancy. The use of an ambiguous term like "a will to live [and] breathe" could hinder collaborative SDM among parents and clinicians by inadvertently placing clinicians' values over the values of parents.

However, if a child did not demonstrate "a will to live [and] breathe" but had a high likelihood of surviving cardiac surgery given their respiratory support status, Rosenblum et al. also state their institution may agree to surgery if the family was committed to the potential

[67] Rosenblum et al., "Cardiac Surgery in Children with Trisomy 13 or Trisomy 18."
[68] Janvier, Farlow, and Wilfond, "The Experience of Families With Children With Trisomy 13 and 18 in Social Networks."

postoperative respiratory support the child needed. They note the value of preoperative discussions and commitments from the family, both of which are goals within a pathways approach to SDM for children with T13 and T18.[69]

Furthermore, one commentator for the paper by Swanson et al. concluded that,

> *"...for those unfortunate children who remain chronically ventilator dependent, it is reasonable to offer palliative care, keeping in mind that what most parents want for their child is the quality of life, not the quantity."*[70]

Similarly, describing children with T13 and T18 who require chronic MV as "unfortunate" is a misguided assumption of their ability and outlook. This determination of what interventions are reasonable relies on a quality-of-life judgment and clinical perception of what parents truly want for their children. Yet parents and clinicians may not share the same goals; some parents may find that full medical interventions, including cardiac surgery, would benefit both the child's quality and quantity of life. Moreover, palliative care and pediatric cardiac surgery are not mutually exclusive; present-day palliative care is about quality of life and living as much as it is about dying, and children with congenital heart anomalies would benefit from an integration of the two specialties.[71]

Indeed, in a parental survey conducted by Davisson et al., all participants reported that they believed their child's quality of life was improved by the treatment their child received, regardless of whether their child was still living or not.[72] These authors noted the importance of

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[69] Andrews et al., "Shared Decision Making and the Pathways Approach in the Prenatal and Postnatal Management of the Trisomy 13 and Trisomy 18 Syndromes."
[70] Minoo N. Kavarana, "Commentary: Cardiac Surgery in Children with Trisomy 13 and Trisomy 18: 'Is It the Quality of Life or the Quantity?,'" *The Journal of Thoracic and Cardiovascular Surgery* 162, no. 1 (July 2021): 252, https://doi.org/10.1016/j.jtcvs.2020.09.027.
[71] Andrea Wan, Kevin Weingarten, and Adam Rapoport, "Palliative Care?! But This Child's Not Dying: The Burgeoning Partnership Between Pediatric Cardiology and Palliative Care," *Canadian Journal of Cardiology* 36, no. 7 (July 1, 2020): 1041–49, https://doi.org/10.1016/j.cjca.2020.04.041.
[72] Davisson et al., "Trisomy 18 and Congenital Heart Disease."

understanding parental perspectives in clinical decision-making as both parents and clinicians pursue an optimal quality of life for the child. Parents with children who have T13 or T18 have described their experiences in detail, with a majority expressing their child's happiness, achievements, positive quality of life, and relationships with their families.[73]

Adopting a role-differentiated approach in Josie's case (Figure 3) highlights why her parents and clinicians may interpret data differently. Her parents and clinicians will value her disability and quality of life in ways the other may not readily understand. Josie's parents will make decisions on her care based on their family values and aspirations for Josie. They may request medical interventions because that is what they believe any good parent would do for a child with special needs. Josie's parents see her potential and love her unconditionally, regardless of her genetic makeup, VSD, reliance on MV, or possibly reduced physical and/or neurodevelopmental abilities. After careful discussion with Josie's medical team, they may understand and be willing to provide the appropriate level of long-term medical care to Josie considering her disabilities. They may disagree with clinicians' quality-of-life judgments because they do not see her condition as unfortunate or an obstacle to her receiving the care a child without T18 would reasonably receive.[74]

Conversely, Josie's clinicians will come to decisions on her care based on their clinical expertise and knowledge of pediatric patients with similar clinical histories. They may be hesitant to recommend or perform surgery to repair Josie's VSD because of her genetic condition and risk factors. For example, Graham et al. and others reported that children with T13 and T18

[73] Janvier, Farlow, and Wilfond, "The Experience of Families With Children With Trisomy 13 and 18 in Social Networks"; Guon et al., "Our Children Are Not a Diagnosis."
[74] Koogler, Wilfond, and Ross, "Lethal Language, Lethal Decisions"; Alaina K. Pyle et al., "Management Options and Parental Voice in the Treatment of Trisomy 13 and 18," *Journal of Perinatology* 38, no. 9 (September 2018): 1135–43, https://doi.org/10.1038/s41372-018-0151-6.

who are unable to self-regulate their breathing preoperatively are historically unable to do so after their operation; therefore, the ability to breathe independently is both a benchmark and barrier to cardiac surgery in these patients.[75] However, a determination of surgical candidacy based on an assumption of normative ability (breathing on one's own) disregards children who will benefit from surgery despite assisted breathing.

If clinicians determine that the burdens of respiratory support outweigh the benefits, it is important they separate medical judgments from quality-of-life judgments.[76] It is well-documented that the term "quality of life," when used by clinicians with respect to children with disabilities, can be a euphemism for their own attitudes towards physical and neurodevelopmental impairments.[77]

Further, in the prenatal and postnatal context for children with disabilities, presumptions about dis/ability are also seen in the language used to describe the quality of life for the family, e.g., language of suffering and pity rather than language of self-determination and support.[78] Similar to the empowerment and respect for self-determination prenatal patients deserve when making choices about pregnancy, parents of children with T13 and T18 should be met with the same respect when pursuing cardiac surgery for their child. SDM is one approach to working

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[75] Graham et al., "Effectiveness of Cardiac Surgery in Trisomies 13 and 18 (from the Pediatric Cardiac Care Consortium)"; David M. Kwiatkowski et al., "Neonatal Congenital Heart Disease Surgical Readiness and Timing," *Pediatrics* 150, no. Supplement 2 (November 1, 2022): e2022056415D, https://doi.org/10.1542/peds.2022-056415D.

[76] Wilfond, "Tracheostomies and Assisted Ventilation in Children With Profound Disabilities."

[77] Koogler, Wilfond, and Ross, "Lethal Language, Lethal Decisions"; Wilfond, "Tracheostomies and Assisted Ventilation in Children With Profound Disabilities"; Shelly Haug et al., "Using Patient-Centered Care After a Prenatal Diagnosis of Trisomy 18 or Trisomy 13: A Review," *JAMA Pediatrics* 171, no. 4 (April 1, 2017): 382–87, https://doi.org/10.1001/jamapediatrics.2016.4798; Pyle et al., "Management Options and Parental Voice in the Treatment of Trisomy 13 and 18"; Tyler Tate, "What We Talk about When We Talk about Pediatric Suffering," *Theoretical Medicine and Bioethics* 41, no. 4 (August 2020): 143–63, https://doi.org/10.1007/s11017-020-09535-8; Michael Kochan et al., "Disagreement About Surgical Intervention in Trisomy 18," *Pediatrics* 147, no. 1 (January 1, 2021): e2020010686, https://doi.org/10.1542/peds.2020-010686.

[78] Sujatha Jesudason and Julia Epstein, "The paradox of disability in abortion debates: bringing the pro-choice and disability rights communities together," *Contraception* 84, no. 6 (December 2011): 541–43, https://doi.org/10.1016/j.contraception.2011.08.022.

through these ethical tensions between parents and clinicians when determining who and what should guide the interventions and goals for a child with T13 or T18.[79]

3.2.2. Quantitative futility and successful surgical outcomes

Futility, much like quality of life, is a complex value judgment that varies based on context and personal values; it is also frequently cited as justification for withholding interventions in children with T13 and T18.[80] Futility arguments can rely on biology and anatomy (physiologic futility), statistics and success rates (quantitative futility), and values-based determinations of worthwhile interventions (qualitative futility).[81] Another focus of this analysis was to identify conclusions associated with quantitative futility or potentially inappropriate interventions given a low probability of a successful outcome.

For example, a care team may determine that cardiac surgery is quantitatively futile for children with T13 and T18 who are receiving MV because of a strong association between this preoperative risk factor and mortality. If an institution considered surgery to be successful only when the child survives to hospital discharge and "preoperative [MV] was associated with significantly increased risk of postoperative mortality," as noted in Cooper et al., then the operation could be considered inappropriate on the likelihood that it fails to reach the desired goal. However, the goals of care will differ based on parents' and clinicians' values, interpretations of "success," and how they understand "significant" statistics.[82]

[79] Andrews et al., "Shared Decision Making and the Pathways Approach in the Prenatal and Postnatal Management of the Trisomy 13 and Trisomy 18 Syndromes."
[80] Koogler, Wilfond, and Ross, "Lethal Language, Lethal Decisions."
[81] Wilfond and Carey, "Parental Requests for Intervention in Children with Lethal Conditions"; Alexander A. Kon, "Futile and Potentially Inappropriate Interventions: Semantics Matter," *Perspectives in Biology and Medicine* 60, no. 3 (2018): 383–89, https://doi.org/10.1353/pbm.2018.0012.
[82] Kon, "Futile and Potentially Inappropriate Interventions."

For the surgical cohort in the Swanson study (N=14), in-hospital mortality was low (N=1; 7%), and they reported a high 1-year survival (70%) and median survival of 3.4 years.[83] In addition, the survival to discharge rate was 75% (N=3) for patients who received preoperative MV (N=4) (Appendix B). Given the small sample size, in-hospital mortality for patients with preoperative MV was 25% (N=1).

The authors did not find nor recommend "that noncardiac comorbidities preclude performing cardiac operations on T13/18 children" and further, advocated for a collaborative clinical assessment that includes, rather than excludes, preoperative and postoperative airway evaluations. Though a majority of children with T13 and T18 in their cohort experienced at least one postoperative airway complication, they did not conclude that cardiac surgery was a futile intervention; rather, they called attention to the importance of communicating perioperative respiratory support and complications with families so they may make informed decisions about the care for their child.

Two commentaries on the Swanson et al. study show how data and interventions can be interpreted as contradictory, even among clinicians.[84] The authors of the commentaries (all physicians) have unique values and experiences that define their role as clinicians. They may value the empirical evidence that while children with T13 and T18 who received cardiac surgery are living longer, high, early mortality rates persist.[85] What is unclear is what mortality data are considered important to determine whether cardiac surgery will be successful in these cohorts. This threshold will vary among institutions, professionals, and parents, as some may have more

[83] Swanson et al., "Impact of Trisomy 13 and 18 on Airway Anomalies and Pulmonary Complications after Cardiac Surgery."
[84] Kavarana, "Commentary"; Douglas M. Overbey, Joseph W. Turek, and Nicholas D. Andersen, "Commentary: Trisomies and Tribulations: Don't Get Blindsided by Pulmonary Complications after Pediatric Heart Surgery," *The Journal of Thoracic and Cardiovascular Surgery* 162, no. 1 (July 2021): 250–51, https://doi.org/10.1016/j.jtcvs.2020.09.052.
[85] Glinianaia et al., "Ten-Year Survival of Children with Trisomy 13 or Trisomy 18."

modest interpretations of that data than others.

Indeed, the author of the first commentary, Kavarana, stated that,

> *"...based on the current, albeit sparse, data, it seems evident that despite the poor long-term survival, low-moderate risk cardiac surgery can be offered to children with trisomy 13 and 18 with a reasonable hospital mortality, provided they are not on a mechanical ventilator preoperatively."*[86]

The authors of the second commentary, Overbey et al., came to a different conclusion from the data, with the authors stating that,

> *"[t]here were signals toward higher rates of tracheostomy and mortality in T13/18 patients that did not reach statistical significance…Overall, this study provides objective data to assist in counseling families before congenital heart surgery in T13/18 patients, and to establish reasonable expectations regarding postoperative recovery and potential long-term complications of surgery, including tracheostomy and ventilator dependence."*[87]

Overbey et al. concurred with Swanson et al., as their institution similarly requires perioperative airway evaluations.[88] Kavarana likewise noted the ethical challenges that arise in this cohort but found low-moderate risk cardiac surgery to be acceptable on the condition that patients were not reliant on MV.[89] Moreover, Kavarana found that there were limited data available to support reasonable in-hospital mortality, but Overbey et al. found that the data presented were objective to guide SDM with parents of children with T13 and T18. These

[86] Kavarana, "Commentary."
[87] Overbey, Turek, and Andersen, "Commentary."
[88] Ibid.
[89] Kavarana, "Commentary."

divergent conclusions show how interventions that qualify as reasonable and worthwhile to one clinician may be regarded as potentially inappropriate and not medically beneficial to another.

These differing interpretations of epidemiological data also show this discordance exists not only between parents and clinicians but within the medical community. Clinical decision-making for children with T13 and T18 exists in a "stable gray zone" where the "right" decision on whether cardiac surgery qualifies as a medically appropriate intervention will undoubtedly vary among the interested parties.[90] The uncertainty as to which interventions are statistically meaningful is due in part to the perspectives of each individual and the values they hold regarding people with disabilities and whether they can "beat the odds."

Additional evidence of how clinicians view the relationship between quantitative futility and successful outcomes is observable in two influential studies: the seminal work from Graham et al. in 2004 and the largest cohort study to date from Cooper et al. in 2019. These authors presented data that support preoperative MV as a significant risk factor associated with postoperative mortality. Graham et al. was the first multicenter study to present postoperative outcomes in children with T13 and T18. They emphasized the effectiveness of cardiac surgery was evident from the fact that all surviving patients who received MV for two days or fewer were discharged without respiratory support, a "stark contrast" to patients who received greater than two days of MV.[91] Similarly in Cooper et al., there was a statistically significant odds of postoperative mortality given MV (unadjusted odds ratios of 8.2 and 8.5 in children with T13 and T18, respectively; both P-values<0.012); they concluded that these data "may be a clinically useful indicator of whether to offer cardiac surgery," suggesting that children with T13 and T18

[90] Lantos, "Trisomy 13 and 18—Treatment Decisions in a Stable Gray Zone."
[91] Graham et al., "Effectiveness of Cardiac Surgery in Trisomies 13 and 18 (from the Pediatric Cardiac Care Consortium)."

who require preoperative MV may not qualify as surgical candidates.[92] Indeed, Rosenblum et al. cited Cooper's study as support for why they preferred to operate on children with "a will to live [and] breathe," perpetuating the notion that cardiac surgery is an inappropriate intervention in children with T13 and T18 who are mechanically ventilated.

From this evidence, one could assume that the clinicians caring for Josie (Figure 3) would rely on these data to explain to Josie's parents why they do not recommend cardiac surgery for her. In their professional opinion, survival statistics for children with T13 and T18 who are preoperatively mechanically ventilated are an obstacle to the desired effect of cardiac surgery: "to provide the safest surgical care and best possible outcomes for every child."[93] As previously noted, the selected epidemiological studies defined operative mortality as within 30 days of surgery or before hospital discharge (Appendix B); this distinction may further indicate cardiac surgery as successful at one institution but not at another. In addition, if Josie's respiratory support status makes her "inoperable," then perhaps it would not be in her best interests to undergo VSD repair at that time. It could be possible that if Josie remained stable on MV, she would become a surgical candidate at a later age when it is safer for her to undergo the operation and the likelihood of her surviving surgery is higher.[94]

But Josie's parents might argue VSD repair is not an inappropriate intervention in light of survival statistics. As her parents, their goals may not be the same as the goals of the clinicians and surgeons. They may assert that the operation is worthwhile if it extends her life, regardless of whether that meets the hospital's definition of success. Whether Josie survives beyond 30 days

[92] Cooper et al., "Cardiac Surgery in Patients With Trisomy 13 and 18."
[93] "Treatments and Services: Heart Surgery," Seattle Children's Hospital, accessed May 22, 2023, https://www.seattlechildrens.org/clinics/heart/treatments-and-services/surgery/.
[94] Davisson et al., "Trisomy 18 and Congenital Heart Disease"; Rosenblum et al., "Cardiac Surgery in Children with Trisomy 13 or Trisomy 18."

post-operation could be arbitrary to them; the more favorable outcome might be taking Josie home, giving her all the love and care they can for as long as they possibly can. Their primary responsibility is to be good parents to Josie, where no intervention is considered futile.

Clinical determinations of futility are individualized based on the goals of care; they can be objective (e.g., it would be physiologically futile to prescribe an antibiotic to treat a viral infection) but in children with T13 and T18, they are more often subjective when based on probabilities and worth. As seen in this analysis, data are not neutral and one's interpretation of evidence is tied to one's values. Consequently, interpretations of morbidity and mortality data can reinforce self-fulfilling prophecies, where cardiac surgery in children with T13 and T18 receiving preoperative MV is repeatedly determined to have a low probability of success.

4. Discussion

4.1. Conclusions

Recent epidemiological data support a growing evidence base that children with T13 and T18, even those who require respiratory support, can successfully undergo cardiac surgery and live longer than current survival estimates. This premise disrupts the once inescapable prediction that life-sustaining interventions were not needed given the high mortality rates in children with T13 and T18.

The studies in this analysis that reported an association between perioperative respiratory support and mortality showed that a majority of patients who received preoperative MV or postoperative tracheostomy placement survived. Framing data in this context may validate claims that cardiac surgery can lead to acceptable survival outcomes in these patients, even with the occurrence of postoperative respiratory complications and management. Furthermore, a role-based approach to this analysis highlights the different but equal merits of parental and clinician

perspectives in pediatric decision-making for children with T13 and T18. Important ethical considerations in ongoing evaluations of cardiac surgery for these children include perceptions of quality of life, dis/ability, quantitative futility, and successful surgical outcomes. Pediatric SDM, including recognition of the equitable weight of parental values, can aid in resolving parent-clinician conflicts on the appropriateness of cardiac surgery in children with T13 and T18 who are also receiving MV.

4.2. Limitations

There are some limitations to this analysis, the first of which was the chosen method for reviewing the literature. Purposeful sampling is not able to identify each epidemiological study that characterized perioperative respiratory support, morbidity, and mortality in patients with T13 and T18. While the literature review for this analysis was structured, conducting a systematic review following the PRISMA checklist could have ensured all literature relevant to the research question was identified.

Next, this ethical analysis did not attempt to address the importance of other sociopolitical factors that influence how medical statistics are interpreted in clinical decision-making. Structural racism, cultural heritage, socioeconomic status, and power dynamics also shape one's understanding of information and can lead to differential treatment and outcomes for children with T13 and T18. Persistent implicit and explicit biases based on race or ethnicity, as well as differing cultural beliefs between clinicians and parents, can play a role in the treatment the child receives and the parent-clinician relationship. Additional influences on the treatment and clinical outcomes for children with T13 and T18 can include knowledge and power imbalances between clinicians and families, and a family's financial constraints to see specialists, access treatment, and maintain long-term care for their child. Further investigation

into the relationship between sociopolitical influences and the interpretation of clinical data will be important in addressing healthcare inequities for children with T13 and T18.

Additional limitations addressed in the retrospective studies include small sample sizes, confounding variables, selection bias, and an inability to comment on long-term outcomes. Few of the selected epidemiological studies, if any, reported 1-year or 5-year survival estimates, and those that did report these data did not differentiate if the survivors received perioperative respiratory support. Further, variations in the accuracy and completeness of hospital reporting of patient deaths could lead to potential misclassification of deaths.

4.3. Looking forward: improving surgical decision-making

A contribution to this field would be an empirical study to assess the current views and rationales of cardiac surgeons regarding surgery in children with T13 and T18 who are receiving MV. A qualitative study using interviews or a survey could highlight the ethical considerations they find most relevant, as well as provide an opportunity for surgeons to explain whether or not they chose to operate on these patients. A national research study could also allow for comparisons of institutional practices and patient outcomes.

In addition, the willingness of cardiac surgeons and their institutions to operate in light of morbidity and mortality data may highlight an underlying social and moral issue in how surgeons perceive dis/ability and make judgments regarding the care of children with T13 and T18. Professional attitudes are a worthwhile factor to explore, particularly if children with T13 and T18 could benefit from cardiac surgery if not for health inequities and implicit disability biases in medicine. This assessment aligns with the SDM approach to counseling families with a child with T13 or T18 who needs cardiac surgery, especially when respiratory anomalies or complications are present. Transparency in surgical decision-making can ensure respiratory

support status is justly considered so these patients may obtain the life-sustaining treatment they deserve.

Appendix A. Cohort characteristics and perioperative respiratory support

First author and year Study	Years of study	Type of study	Cohort	No. of patients	Cardiac surgery operations	Primary cardiac operation	Persistent pre-operative hospital-ization	No. of patients with preoperative respiratory support			No. of patients with postoperative respiratory support		
								Pre-operative MV	Pre-operative trache-ostomy	Pre-operative HFNC (>1 L/m)	Post-operative MV	Post-operative trache-ostomy	Post-operative HFNC (>1 L/m)
Graham EM 2004 "Effectiveness of cardiac surgery in trisomies 13 and 18 (from the Pediatric Cardiac Care Consortium)"	1982-2000	Multicenter retrospective cohort	T13/T18	35	35	VSD (20) TOF (6) Other (9)	n/a	9 (>2 days)	n/a	n/a	5	n/a	n/a
								26 (0-2 days)	n/a	n/a	0	n/a	n/a
Davisson NA 2018 "Trisomy 18 and Congenital Heart Disease: Single-Center Review of Outcomes and Parental Perspectives"	2005-2017	Single-center retrospective cohort	T18	17	9	VSD (7) TOF (1) Other (1)	n/a	3	n/a	n/a	n/a	5	n/a
Cooper DS 2019 "Cardiac Surgery in Patients With Trisomy 13 and 18: An Analysis of The Society of Thoracic Surgeons Congenital Heart Surgery Database"	2010-2017	Multicenter retrospective cohort	T13	304	73	VSD (103) VSD/CoA (9) TOF (24) Other (207)	n/a	16	n/a	n/a	n/a	n/a	n/a
			T18		270			82	n/a	n/a	n/a	n/a	n/a
Swanson SK 2020 "Impact of trisomy 13 and 18 on airway anomalies and pulmonary complications after cardiac surgery"	1994-2014	Single-center retrospective case-control	T13/T18	14	14	VSD (9) VSD/CoA (2) TOD (2) Other (1)	9	4	0	1	7	2	2
Rosenblum JM 2022 "Cardiac surgery in children with trisomy 13 or trisomy 18: How safe is it?"	2002-2021	Single-center retrospective cohort	T13/T18	19	19	VSD (8) TOF (7) Other (4)	5	0	2	6	n/a	n/a	n/a

MV (mechanical ventilation); HFNC (high-flow nasal cannula); VSD (ventricular septal defect); TOF (Tetralogy of Fallot); CoA (coarctation of the aorta); Other (may include patent ductus arteriosus closure, atrial septal defect repair, pulmonary artery banding, pulmonary valve reconstruction, and/or Blalock-Taussig shunt)
n/a. Data characterizing perioperative respiratory support are not available.

Appendix B. Association between perioperative respiratory support and survival

First author and year Study	Years of study	Type of study	Cohort	Preoperative MV				Postoperative tracheostomy			
				No. of patients with preoperative MV	Survival to discharge No. (%)	In-hospital mortality within 30 days of surgery No. (%)	In-hospital mortality before discharge No. (%)	No. of patients with postoperative tracheostomy	Survival to discharge No. (%)	In-hospital mortality within 30 days of surgery No. (%)	In-hospital mortality before discharge No. (%)
Graham EM 2004 "Effectiveness of cardiac surgery in trisomies 13 and 18 (from the Pediatric Cardiac Care Consortium)"	1982-2000	Multicenter retrospective cohort	T13/T18	9 (>2 days)	8 (89%)	1 (11%)	0 (0%)	n/a	n/a	n/a	n/a
				26 (0-2 days)	24 (92%)	2 (8%)	0 (0%)	n/a	n/a	n/a	n/a
Davisson NA 2018 "Trisomy 18 and Congenital Heart Disease: Single-Center Review of Outcomes and Parental Perspectives"	2005-2017	Single-center retrospective cohort	T18	3	2 (67%)	0 (0%)	1 (33%)	5	4 (80%)	0 (0%)	1 (20%)a
Cooper DS 2019 "Cardiac Surgery in Patients With Trisomy 13 and 18: An Analysis of The Society of Thoracic Surgeons Congenital Heart Surgery Database"	2010-2017	Multicenter retrospective cohort	T13	16	11 (69%)	5 (31%)b		n/a	n/a	n/a	n/a
			T18	82	52 (63%)	30 (37%)b		n/a	n/a	n/a	n/a
Swanson SK 2020 "Impact of trisomy 13 and 18 on airway anomalies and pulmonary complications after cardiac surgery"	1994-2014	Single-center retrospective case-control	T13/T18	4	3 (75%)	1 (25%)	0 (0%)	2	2 (100%)	0 (0%)	0 (0%)
Rosenblum JM 2022 "Cardiac surgery in children with trisomy 13 or trisomy 18: How safe is it?"	2002-2021	Single-center retrospective cohort	T13/T18	0	n/a	n/a	n/a	n/a	n/a	n/a	n/a

MV (mechanical ventilation)
n/a. Data characterizing survival as a function of perioperative respiratory support are not available.
a. Patient also received preoperative mechanical ventilation
b. Data to differentiate in-hospital mortality are not available