Ethical Challenges Posed by Trisomy 18 Infants

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The prognosis for newborns with full trisomy 18 is very poor, with approximately 10% surviving to one year [1]. Roughly 60% of infants born in the United States with trisomy 18 do not survive to hospital discharge [2]. Although there are rare reports of long-term survivors, advances in medical technology over the past 20 years have not had an appreciable effect on survival of these infants [3].

Prenatal diagnosis of this syndrome offers women several choices: they can choose to terminate the pregnancy, carry the child to full term followed by palliative care, provide supportive care for the infant (oxygen, breathing assistance, artificial feeding, etc.) and take them home or place them in a hospice, or provide aggressive treatment including surgical repair of cardiac anomalies [4]. Because of significant advances in neonatal technology and operative procedures, the range of options available to parents who choose to attempt to prolong their trisomy 18 infant's survival aggressively has increased greatly. Furthermore, the willingness of surgeons to intervene in selective cases has also increased.

Given the increasing number of options open to parents with trisomy 18 babies, ethical issues have arisen which can best be summed up by asking the question: just because we *can* intervene in these ways, *should* we do so? What are the appropriate ethical criteria to justify treating or not treating trisomy 18 babies with aggressive life-prolonging procedures or operations? These questions are increasingly asked across the medical decision-making spectrum as technology progresses. In particular, end-of-life decisions involving similar ethical considerations occur daily in all hospitals.

In this issue of *IMAJ* the article by Toker and Salzer [5] asks "how far should we go?" in treating newborns with trisomy 18. They present the case of an infant with this condition born to an ultra-Orthodox Jewish couple who refused amniocentesis despite knowing that their child would be born with a cardiac malformation and additional structural defects. Whether their decision to decline amniocentesis was due to religious considerations that would prohibit termination is not mentioned. The diagnosis of trisomy 18 was made shortly after birth, and a meeting of the local ethics committee was held with the parents present to discuss treatment options. The authors state: "After a long debate the ethics committee recommended that the medical teams perform heart surgery on the baby, mainly due to updated literature that described better prognosis for babies with trisomy 18 who underwent heart surgery."

Details of the pro and con arguments at that crucial meeting are understandably not presented in the article, but we are left with critically important unanswered questions that impact heavily on the ethics of the case. Any such decision relies heavily on a cost-benefit assessment with many issues on both sides. To mention a few: how much "better" would the infant's prognosis be in terms of quality and quantity of life with an operative approach: might the child live an additional 6 months? A year? More? How might other clinical considerations such as improved quality of life or decreased need for hospitalization affect the applicability of the data in the two studies cited by the authors that justified the operative approach? How medically sophisticated were the parents? What were their goals for their child? Would they choose to extend life at all costs? How suited were they to care for this baby if she survived? Were there other children in the family? What about issues of home resources, both financial and emotional? Would additional surgeries be necessary? Would the infant undergoing these interventions suffer unduly? To what extent did their religious beliefs influence this decision?

Unfortunately, the infant required a total of 5½ weeks on a ventilator after heart surgery. She was diagnosed with a fatal liver condition – biliary atresia – a few months after surgery and she died a little over 5 months of age. A second ethics committee meeting was held shortly before the infant's death and determined that all possible further treatments would be "futile." Supportive care was therefore recommended. She spent 2 months of her short life at home and was hospitalized for the rest.

The authors correctly point out that infants with severe chromosomal abnormalities pose the greatest ethical dilemmas to clinicians who, along with the parents, must grapple with the question of how much treatment to provide. The question is more acute today because of the vastly increased

number of medical and surgical options available to treat these infants. Issues of quality of life, patient and family suffering, and appropriate resource allocation are uppermost in these decisions.

When considering these dilemmas, we do not think it helpful to invoke the issue of futility – a strong term that implies that the intervention under consideration should not be carried out – unless the specific goal(s) that cannot be achieved are specified. A treatment that may be futile in one case is not futile in another with different goals of care. The authors state that in cases of infants with severe chromosomal abnormalities, "treatment is usually futile… ." If the goal of treatment is to prolong the life of an infant long enough to enable the child to be cared for at home for several months or longer, the treatment being considered may be anything but futile. If the goal is to prolong the child's life by years or to improve his or her mental development, then the treatment in question might well be futile. Unless one states the goals of treatment, the futility or efficacy of an intervention cannot be assessed. Without specification, one is left to wonder if the heart of the issue is whether the treatment is futile in terms of prolonging life or improving the quality of life.

Another term that is probably best avoided is "lethal." The implication of the term is that there is no treatment available to significantly prolong life. Thus, deeming a patient's condition lethal allows the physician to not offer any treatment other than comfort care. For conditions such as trisomy 18, trump cards such as the word "lethal" serve as thinly veiled quality of life judgments by those opposed to treatment. Arguments grounded in the anticipated burdens and benefits of a possible treatment will carry greater moral weight than any label [6].

We agree with the authors that "the role of the ethics committee is crucial" in cases such as this. But we would not limit the committee's responsibility "to resolve and clarify any conflicts between parents and physicians and among physicians." An ethics consultation in such a case can be most useful in clarifying the issues involved: what are the parents' goals for their child? Are these goals realistic? Have the parents' financial and emotional resources been duly considered by the medical team before reaching a conclusion? Have the physicians done due diligence in collecting current data that will enable the parents to make an informed decision? Has there been good communication between the parents and physicians? Have the religious needs of the parents been dealt with? Presumably if these and other questions have been adequately addressed, the best interests of the baby will have been served as well.

The authors correctly state that physicians and ethics committees are often faced with the dilemma of having to make recommendations and decisions based on inadequate data because of the rapid pace of technological advances. In such cases, perhaps having more than five members on the ethics committee might be helpful to include as broad a representation of disciplines and expertise as possible. Although we do not profess to have the ideal formula for ethics committee representation, we do have a much larger pediatrics ethics committee than the authors do. Our committee has 25 members representing medicine, nursing, social work, law, psychiatry, pastoral care and hospital administration, as well as community representatives. The physicians on our committee represent neonatology, pediatric critical care, psychiatry, neurology, cardiology, palliative care, and oncology. Consults are conducted by small teams of ethics committee members (usually three members) who almost always meet with the family as well as with the nurses and physicians treating the patient. All consults are ultimately reviewed by the full committee.

Finally, we question the conclusion of the authors that the best case scenario in such cases is "to get parental consent for the committee's decision." This sounds overly authoritarian. Much has been written over decades about what constitutes an ethics consultation. A simple but useful definition might be "a service provided by an individual ethics consultant, ethics consultation team or ethics committee to help patients, providers, and other parties resolve ethical concerns in a health care setting" [7]. We therefore suggest that the goal of the ethics committee deliberation in this case should be to involve participants in such a way that the decision for further treatment of the child represents as much as possible a convergence of views between staff and family based on the rights, values and ethical concerns of the family and the medical staff within a context of societal values, laws, and institutional policies. In general, a consensus may not always be realized, but we believe that assiduous efforts on the part of the ethics consultants to foster better communication between families and staff and to educate families about the medical issues will usually result in a treatment plan that is ethically and medically appropriate. When such resolution is not possible, further legal or administrative measures may have to be taken.

The challenges of treating a trisomy 18 infant are a paradigm for other ethical conundrums that will continue to arise in medicine due to striking advances in medical technology. Although not without their detractors [8], medical ethics consultants and committees who carry out their tasks wisely and sensitively can serve as useful advisors and mediators to patients, families and medical professionals.

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