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Original Article

Informed consent, *bioethical equipoise*, and hypoplastic left heart syndrome

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Abstract In utero diagnosis of complex progressive cardiac disease such as hypoplastic left heart syndrome presents a novel opportunity for antepartum, intrapartum, and neonatal management. The clinical possibilities and potential for differing outcomes challenge the mother–foetus dyad with regard to informed consent. Previous studies reveal that rates of termination of pregnancy for foetuses with hypoplastic left heart syndrome vary widely in the United States and Europe, leading us to surmise that informed consent may be practised differently. The purpose of this paper is to review the ethical considerations and physician responsibilities of informed consent as they relate to prenatal and postnatal patients with hypoplastic left heart syndrome. Special consideration is paid to the informed consent process as practised by the obstetrician, perinatologist, paediatric cardiologist, and paediatric cardiac surgeon as it relates to termination of pregnancy, comfort care, and surgical palliation. We will argue that informed consent as it relates to hypoplastic left heart syndrome is far from standardised and that there exists a state of *bioethical equipoise* concerning the extent and limits of its application in the current clinical setting.

Keywords: Norwood; single-ventricle repair; pregnancy termination; comfort care; surgical palliation; foetal diagnosis

Advances relating to in utero diagnostics have dramatically altered the identification and management of complicated pregnancies.[1] At the same time, these advances also present new challenges to patient care for both the mother and the developing foetus. For instance, the in utero diagnosis of complex progressive cardiac defects such as hypoplastic left heart syndrome presents a novel opportunity for antepartum, intrapartum, and neonatal management in ways that could not have been imagined otherwise. Yet, innovation such as this invokes a myriad of questions and debates about the maternal–foetal dyad, specifically those relating to interests of the mother and foetus, as well as disciplinary biases that different clinicians bring to light.[2] Ultimately, many of these discussions are centred on the issues of patient autonomy, decision making, and informed consent. Thus, as medical innovation in the field of obstetrics and neonatology advance, it will be critical to consider informed consent and the vital role it plays in contemporary medicine.

Many authors instantiate informed consent into its legal meaning. In this case, informed consent is most commonly used to refer to a series of legal precedents[3] that prescribe acceptable and reasonable behaviours for physicians and other healthcare professionals in their interactions with patients. The term "informed consent" was initially coined in 1957 as a result of *Salgo v. Leland Stanford Jr*

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University Board of Trustees[4] in which a California court ruled that "a physician violates his duty to his patient and subjects himself to liability if he withholds any facts which are necessary to form the basis of an intelligent consent by the patient to the proposed treatment". Since that time, there has been an ongoing evolution of the legal concept of informed consent with a move from professional standards to patient-oriented standards. In this way, informed consent has been framed in a way that prioritises the patients and their autonomy as the seat of medical decision-making.

In the clinical setting, the term informed consent is most commonly associated with its legal concepts. Yet, its ethical underpinnings reflect the broader meanings of informed consent. At its core, informed consent is a discussion between patients and their healthcare provider. The patient's autonomy and right of self-determination are prioritised in the decision-making process, allowing them to make medical decisions that reflect their beliefs and healthcare needs.

The purpose of this paper is to review ethical considerations and physician responsibilities of informed consent as they relate to prenatal and postnatal patients with hypoplastic left heart syndrome. We will argue that informed consent as it relates to hypoplastic left heart syndrome is far from standardised and that there exists a state of *bioethical equipoise* concerning the extent and limits of its application in the clinical setting.

Clinical background

Before the 1990s, patients born with hypoplastic left heart syndrome died shortly after birth owing to maldistribution of systemic and pulmonary blood flow, patent ductus arteriosus closure, and circulatory collapse. The introduction of the Norwood procedure[5] made it possible for these infants to undergo staged palliation, which culminated in some variation of the Fontan operation – single-ventricle repair. Shortly thereafter and largely resulting from the high mortality of the Norwood operation, Bailey,[6] Mavroudis,[7] and Idriss[8] applied neonatal orthotopic cardiac transplantation to these patients in an effort to provide a two-ventricle solution, albeit with lifelong antirejection medications and, at the time, unknown long-term complications. Given the limited number of therapeutic options during that era, the informed consent discussion with new parents addressed three primary choices available for newborns with this condition: the Norwood operation, cardiac transplantation, and comfort care. In time, the evolving improvement in outcomes after the Norwood procedure in contrast to the difficulty of purveying donor hearts in a timely manner shifted the therapeutic paradigm towards staged palliation, reserving cardiac transplantation for patients who failed the Norwood pathway.

During this metamorphosis, researchers at Boston Children's Hospital[9] introduced a new way to address issues of hypoplastic left heart syndrome in utero using a foetal catheter intervention for two cohorts of foetuses with hypoplastic left heart syndrome: foetuses with severe aortic stenosis/small ventricle and foetuses with intact or nearly intact atrial septum. This marked the beginning of in utero procedures for these conditions. These procedures require a large team of specialists to provide intra-operative support for both the mother, including specific obstetrical management to prevent uterine contractions during the procedure, and the foetus. Once the gravid woman is stable and has sufficient anaesthesia, a catheter can then be introduced through the skin, uterus, chest cavity of the foetus, and the apex of the left ventricle or the right atrium, respectively.[10]

The rationale behind foetal transventricular aortic valve dilatation is to increase flow through the left side of the heart and allow foetal left ventricular growth in utero, resulting in biventricular physiology.[11] The rationale for transatrial septostomy is to decompress the left side of the foetal heart and reverse the severe pulmonary vascular disease associated with pulmonary venous obstruction secondary to intact or nearly intact atrial septum. This procedure will facilitate and enhance the outcomes associated with the Norwood operation after birth. The programme is still largely experimental and the results so far have been mixed.[12]

Special considerations of prenatal and postnatal informed consent for hypoplastic left heart syndrome

Advances in foetal echocardiography have greatly enhanced the ability of parents to know, in advance, and with a high degree of certainty of their future child's diagnosis.[13] The informed consent process should recognise and respect the pregnant woman's autonomy, wishes, values, and beliefs in the context of sensitivity and empathy. To make the process more complex, there are a number of specialists (physicians) who interact with the mother at different stages during the pregnancy depending on her wishes. Most reports with regard to informed consent in this setting centre on the foetus and potential outcomes if the foetus is carried to term. What is generally not reported is the informed consent process involving the mother in the event that she chooses to terminate the pregnancy. Although specialised clinicians take the lead in each situation – obstetrician or perinatologist for referrals or termination of pregnancy; paediatric cardiologists,

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paediatric cardiac surgeon for foetus management after birth – there is one pregnant patient with concerned family members who is confronted with all the health issues at hand. All clinicians must remember this and acquire an appropriate amount of information in order to facilitate informed consent consistent with the number of attending physicians.

In order to make an informed decision, pregnant women must comprehend the medical facts about the foetus. This information includes data about surgical outcomes at their respective institution, other institutions, and the different operative procedures that might be necessary. Future considerations include the possibility of neurological deficits, expected longevity, and possibility of eventual cardiac transplantation. Informed decisions also entail consideration of one's values and beliefs and making a choice regarding the medical facts that is internally consistent with those values and beliefs.[14] Patients also need to have the opportunity to openly discuss their thoughts and opinions about giving birth and caring for a child with these specific healthcare needs, specifically issues of comfort care in the context of social acceptance, family unity, and economic accountability. In doing so, they must also be informed of the option of termination of pregnancy and considerations for future childbearing while being given sufficient opportunity to reflect on these considerations.

Given the magnitude of such decisions, it is unrealistic to think that the decision-making process about continuing or ending a pregnancy can be initiated and concluded in the time span of a single meeting.[3] Fostering a proper understanding of the many complex issues involved in the anatomy, physiology, prognosis, treatment options, and long-term expectations is a herculean task, which requires time for both physicians and patients to grasp the range of intellectual, emotional, and ethical challenges that accompany each diagnosis. It may take multiple visits to slowly and conscientiously educate the parents without undue pressure and pessimism. It may be necessary for the clinician to find the "Golden Mean"[15] in the informed consent process, which may be different for different families. However, a balance must be found in these discussions. Timing is everything in pregnancy; each delay in decision-making brings the patient closer to the time of viability, placing important limitations on both the decision to end the pregnancy but also to perform in utero procedures to manage the condition.

Presently, the informed consent process is in evolution as the clinical management of patients with hypoplastic left heart syndrome changes in league with advances in technology. Presumably, advising clinicians discuss termination of pregnancy, the three operation Norwood pathway, and comfort care after birth. Dialogues are then established where sensitive issues can be discussed depending on the course of the conversation such as local and distant surgical outcomes, the physical limits and longevity of the Fontan operation,[16–18] and eventual cardiac transplantation with the attendant lifelong immunosuppression regimens. Owing to the fact that the range of topics discussed in obtaining informed consent for these patients is not standardised,[14] there can be no certainty as to whether or not all of these important issues are being discussed throughout the field. As such, the experience that physicians and other healthcare providers bring to the conversation has unequal weights throughout the institutions in which they are performed. For instance, the pregnant women in these scenarios will most likely encounter the medical opinions of an obstetrician and a paediatrician, each with different perspectives on how to weigh the medical priorities of the maternal–foetal dyad. Therefore, it can become difficult for physicians and for patients to comprehend fully the reported outcomes of different institutions, as these can be a function of the experience of the individual institution with the proposed procedure. What is more, the physician bias is not the only confounding source in counselling patients and in reporting outcomes. Patients bring with them their own baseline health literacy and ways of acquiring medical information in addition to ideas from their cultural background, their religious beliefs, their baseline knowledge of the disease process, and their values and beliefs about the pregnancy.[19,20] The resulting permutations and combinations of physician and patient points of view create a myriad of different scenarios, each of which necessitate a unique informed consent discussion that meets the informational needs of the patient and the situation. For example, the presentation of informed consent may be different for the hypothetical hypoplastic left heart syndrome foetus with obstructed pulmonary veins and/or associated non-cardiac lesions for a mother with four healthy children at home as opposed to the hypothetical foetus without genetic or associated lesions for a mother with no children who has been trying to get pregnant for a number of years. Clearly, there cannot be a different model of informed consent discussion for each unique set of variables; many would argue that the ability to adapt one's discussion to fit within the given situation is part of the "art of medicine".[14] The challenge, then, becomes balancing one's ethical obligation for full disclosure, with the need to remain sensitive to each individual patient's needs and beliefs.

Clinicians in some programmes[21,22] cite increased Norwood survival as evidence to offer the operation

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without discussing comfort care. They cite other conditions such as prematurity at 24 weeks, other complex forms of challenging congenital cardiac defects, and serious cancer therapies that have not been part of the dialogue of comfort care, despite the enormous costs, challenging neurological outcomes, and as yet unknown long-term outcomes associated with these therapies.[23] Informed consent therefore, in these programmes, means informing the family of the three-staged operations and the attendant complications associated therewith. It is unclear whether cardiac transplantation and lifelong immunosuppression is mentioned under these circumstances. Of course, clinicians should answer questions concerning alternative measures, including comfort care, and are duty bound to refer the family to another institution for comfort care if they are against the idea and feel uncomfortable with its implementation – conscientious objection.[24] It is also unclear whether during the informed consent process, clinicians are duty bound to inform the family of the experimental foetal catheterisation programme, regardless of whether they endorse the programme. Some researchers[25] contend that the referring clinician is duty bound to volunteer this information and assist the family in obtaining a second opinion if they desire. How the clinicians inform the family of their recommendations and their biases is not clear, although several ethicists have opined that physicians should share their thoughts with the family and offer a recommendation at an appropriate time.[3]

Who is responsible for informed consent?

We have, thus far, referred to the physician who is responsible for informed consent as, "the clinician". In fact, there are a number of physicians who care or will care for the mother and foetus with hypoplastic left heart syndrome in the structure of team care and group practice. In general, it is the obstetrician who performs a preliminary evaluation of the foetal heart in high-risk parturients. In many cases, abnormal findings will lead to a referral to a perinatologist who conducts comprehensive foetal screening and will identify abnormal foetal cardiac anatomy. After a discussion with the mother, the perinatologist may elect to have an informed discussion about all available options. One option would be to continue management with the intention of carrying the pregnancy to term. In these cases, the perinatologist would refer the mother to a paediatric cardiologist who would perform a comprehensive foetal echocardiogram and engage in the informed consent process that includes the options that have been noted in the aforementioned discussion. In the United States, the perceived standard of care has been that comprehensive informed consent is given by the paediatric cardiologist.[26] This will then result in the options of termination of pregnancy or continuation of pregnancy to surgical therapy or comfort care. We recognise that a pregnant woman may make the choice to end a pregnancy when she can make an informed decision. However, we recommend that consultation with a specialist is a core component of the balanced counselling process. In this way, the patient can be prepared to consider the advantages and disadvantages of continuing or terminating the pregnancy. In the event that termination is elected, it will be her obstetrician who will have an informed discussion with her about the procedures involved, considerations of risk/benefit, and future reproduction issues.

If the mother decides on continuing the pregnancy with the intent of staged intervention or transplantation, then a referral is made to the surgeon for further informed discussions concerning the surgical procedures, complications, and the like.[25,27] By the multi-disciplinary nature of these complex cases, the informed consent discussion becomes inclusive of several members of the healthcare team to take advantage of the involved physicians with their particular perspective and expertise.

What actually happens in the informed consent process from the obstetrician to the perinatologist to the paediatric cardiologist to the surgeon is presently unknown.

Standardised informed consent?

The preceding discussion has demonstrated the great variability inherent in the informed consent process. This variability includes both heterogeneous content of the discussion *and* the variety of healthcare professionals conducting the consent process for pregnant patients who have a foetus with hypoplastic left heart syndrome. The question then becomes whether or not informed consent needs to be standardised. And if so, what are the tenets of the process? What clinical and social data should be discussed? How do patients make decisions regarding this information in the context of hypoplastic left heart syndrome? There are differences in hypoplastic left heart syndrome patients with associated lesions such as obstructed pulmonary veins, genetic syndromes, and neurological challenges that negatively influence their clinical outcomes.[28] These conditions are associated with impaired psychosocial development.[29] There are data to show that there is an increased divorce rate in the parents of congenital cardiac surgery patients.[30] Brosig et al[31] reported that parents of children with hypoplastic left heart

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syndrome experienced a more negative impact of the child's illness on the family and more parental stress than the comparative group of parents of children with transposition of the great arteries. And even in the best of circumstances,[32] Fontan physiology limits the patient's exercise tolerance, to say nothing about the high propensity of atrial arrhythmias, and the occasional but lethal consequences of protein-losing enteropathy and plastic bronchitis. Alongside this pessimism is the virtue of hope.[33] Although it is common for families to hope for the best possible outcome, what is the responsibility of the informing physician with respect to the hopes of a family who must make key decisions in the face of considerable uncertainty? Citing figures concerning short- and intermediate-term survival is a possibility, although it is in no way indicative or predictive of what will occur with respect to a given individual patient. As importantly, the entire history of successful surgical therapy for hypoplastic left heart syndrome is a sufficiently recent phenomenon, and no one can speak knowledgeably about true long-term outcomes – of three decades or more. Should the emphasis of counselling be on the very real possibility of survival with the potential for physical or neuro-developmental outcomes that are different from those of individuals without hypoplastic left heart syndrome? Or is it more appropriate to stress the positive aspects of potential outcomes? This relates partly to the optimist's claim that, "Between now and then, who knows what will happen?" When surgeons engaged in the process of informed consent before performing first stage Norwood procedures or neonatal cardiac transplantation two decades ago, they could say little more about potential outcomes than to report the fact that death without surgical therapy was a virtual certainty. At present, discussion of possible outcomes includes not just the potential for serious adverse outcomes as enumerated above, but the fact that among the survivors of surgical management of hypoplastic left heart syndrome there are Eagle Scouts,[34] college students,[35] and two young women who 20 and 23 years after their own initial Norwood operations gave birth to infants with structurally normal hearts.[36] "Between now and then, who knows what will happen", is a driving force behind many of the innovations that extend survival in patients with a variety of diseases. Is it compassionate, generous, even obligatory, or alternatively unfair or self-serving to nurture the optimism of patient families by sharing reports of the best possible outcomes to date?

In reality, there are research protocols that envision improvement of ventricular function through stem cell research.[37] Foetal surgery is possible and may find a niche in the surgical therapy of hypoplastic left heart syndrome.[25,27] Extracorporeal right-sided pumps may serve as destination therapy and improve the hepatic and renal function in failing Fontan patients,[38] and there is always the elusive key to solving the immunologic basis of organ rejection.[39] Yet upon hearing that xenotransplantation is a modality for the future, Shumway remarked, "xenotransplantation is the future of transplantation … and will always be".[40] There is some truth characterised by the limits of our possibilities. Although we have made great strides in cancer therapy, we have not cured all cancers, we have not cured arthritis, and we have not made progress to define and ameliorate ageing. Will these problems ever be solved?

A management paradigm in flux

The range of clinical options presented to families with a diagnosis of hypoplastic left heart syndrome has been addressed in the literature, and there are strikingly different trends in clinical practice between North America and Europe.[41] In North America, there has been a gradual decrease in termination of pregnancy and comfort care for foetuses and neonates, respectively, with hypoplastic left heart syndrome owing to increased survival with the Norwood operation.[42] The rate of termination of pregnancy for hypoplastic left heart syndrome in the United States, taking into consideration multiple sources,[23] is roughly computed to be about 20%. In Europe, the number of neonates born with hypoplastic left heart syndrome is a fraction of what is seen in North America because of the advent of foetal echocardiography and termination of pregnancy.[41] Khoshnood et al[43] reported on the trends in prenatal diagnosis, termination of pregnancy, and perinatal mortality of newborns with congenital cardiac disease in France (1983–2000). During this time, France pursued an active policy of antenatal foetal echocardiography surveillance. They found that almost 90% of the cases of hypoplastic left heart syndrome were diagnosed prenatally and that 60% were terminated before birth. Other European countries have experienced similar results.[44–46]

Reasons for this apparent discrepancy are largely speculative. The influence of local cultural beliefs, values about termination of pregnancy, and raising a child with a potential serious medical condition or disability may be important factors in the decision of whether or not to terminate a pregnancy. Different healthcare systems may also impose tacit restrictions on healthcare delivery and are important considerations in this discussion. These issues, however, are beyond the scope of this paper.

A unified bioethical policy on this issue is very problematic as noted in the aforementioned discussion.

What are the ethical frameworks for recommending termination of pregnancy to a woman when alternative management options exit? What about comfort care in the setting of hopeless conditions? In these cases, what is the role of inducing pregnancies pre-term when the outcomes are expected to be especially poor? Many clinicians would recommend comfort care for patients with trisomy 18 or trisomy 13, although there is some controversy in this regard.[47,48] Of course these examples represent the extreme end of the spectrum. Conversely, there are many clinicians who might have difficulty with the notion of terminating a pregnancy for a foetus with tetralogy of Fallot. Pertaining to foetuses with hypoplastic left heart syndrome, is there a subgroup so challenged by existing therapeutic interventions that would lead clinicians to consider informed consent skewed towards termination of pregnancy or comfort care? Many clinicians would not recommend surgery for hypoplastic left heart syndrome and obstructed pulmonary venous return in a term infant. Similarly, various genetic syndromes might skew clinicians to avoid palliative surgical intervention, especially if the immediate and long-term outcomes are significantly challenged. These trends have occurred many times in the course of other therapeutic indications.

To be sure, informed consent is very patient specific with a complex interaction between the reality of science and the virtue of hope. There is a myriad of contrasting, interactive cultural and religious traditions. In particular, religion is rarely mentioned in medical manuscripts, but it is an important influence on family decision-making; a child is seen as a gift of God.[41] And yet there are things that we have learnt that beckon us to a thoughtful pause concerning hypoplastic left heart syndrome with associated genetic lesions, closing atrial septum, obstructed pulmonary veins, extra-cardiac lesions, and challenging social issues.

Inferences and *bioethical equipoise*

We have shown in this manuscript that there are fundamental differences in the way that hypoplastic left heart syndrome is managed between Europe and North America. Although we do not know many of the details, it appears that the practice of informed consent may be central to these observed differences and that at the present time there is no coordinated method of studying informed consent. This dialogue is largely seen as an offshoot of the physician–patient relationship, which is thought to be personal, hopeful, pragmatic, cultural, and complex. Complicating this issue is the apparent reality that informed consent is rarely taught to students and residents apart from the importance of patient autonomy, non-malfeasance, beneficence, and justice that is emphasised in medical school and in yearly courses in medical institutions. The idea of full disclosure in the face of a terrified family who has just been told of their baby's (foetus's) health challenges seem very draconian and represents the antithesis of what we as physicians are trained to do. This begs the questions of, "Can informed consent be taught?" and "Can informed consent be studied?" We can create "laundry lists" of what should be included in any discussion involving a disease process to make sure that the essential aspects of diagnosis, treatment, and potential complications/death are discussed in some manner that takes into consideration cultural awareness, patient understanding, and fulfillment of expectations. It appears that these tenets are practised differently in several institutions in North America and Europe, which leads us to the conclusion that there is *bioethical equipoise* in the practice of informed consent for hypoplastic left heart syndrome. Benjamin Freedman[49] wrote, "The ethics of clinical research requires equipoise – a state of genuine uncertainty on the part of the clinical investigator regarding the comparative therapeutic merits of each arm in a trial". However, in his manuscript, he further dilates on this issue pointing out that it is actually quite unusual for an individual clinician/investigator to be entirely without bias or experience-based beliefs regarding the relative merit of two treatment strategies, which may be treatment arms of a trial. He then suggests an "alternative concept of equipoise", which would be based on present or imminent controversy in the clinical community over the preferred treatment. According to this concept of clinical equipoise, the requirement is satisfied if there is genuine uncertainty within the expert medical community – not necessarily on the part of the individual investigator – about the preferred treatment. It follows therefore that there is *bioethical equipoise* as it relates to informed consent for hypoplastic left heart syndrome and is based on the idea that there is "controversy in the clinical community over the preferred" method of informed consent, which clearly yields two different pathways in clinical outcomes between North America and Europe. Clinical equipoise is more easily defined and characterised because it usually involves a dyad of medical (drug trial) or surgical treatment patterns. A randomised trial is planned, executed, and oftentimes resolves the controversy.[50] *Bioethical equipoise*, however, is a state of controversy and uncertainty that is grounded in a complex set of factors, including consideration of all the issues raised in this manuscript. In order to study the problem of *bioethical equipoise*, study methods are required with important long-term outcome measurements such as

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the comprehensiveness of informed consent, patient understanding and awareness, realisation of expectations, impact on the family structure, hardship on the patient, and the difficult subject of societal resource allocation.

Clearly, this is no easy task. The kinds of resources required to create such multi-centre studies used to rectify clinical equipoise are immense and are based on the premise of offering a direct, measurable improvement in patient care. Metrics to measure patient satisfaction, decision-making, level of patient education, and amount of disclosure seem cumbersome owing to the subjective nature of the study. Moreover, what value do we, as a society, place on such metrics? No one would argue against evaluating primary outcomes such as death, presence or absence of symptoms, and other measures of clinical treatment success. Psychosocial factors such as those mentioned in this paper with regard to informed consent are not well represented in multi-centre trials, which may indicate that these factors are less understood, less easily ascertained, or not as highly valued in medicine as a whole. The problem of informed consent plagues the clinical management of patients with hypoplastic left heart syndrome perhaps more so than other clinical scenarios in other specialities, as there is more uncertainty inherent in the diagnosis, in the physician–patient(s) relationship, and in long-term outcomes. As such, more attention should be paid to resolving the current *bioethical equipoise* in which providers and patients find themselves. Whether this means multi-centre trials, formulating checklists for topics to be discussed, or standardising the language used in such conversations, this equipoise needs to be addressed. The medical problems that these patients face are troubling enough without the added uncertainties raised by differing levels of disclosure in the informed consent consultation visit. Our duty as physicians is to minimise suffering for these patients and their families. Although we cannot account for all possible differences in patient perspectives and backgrounds, we can simplify a very emotional and difficult process of informed consent for patients by coming to an agreement on which options to discuss.

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