Volume 1 • Issue 1 • 2017

Editorial

Therapeutic Advances in Cardiology

ISSN: 2575-5161

Ethical Issues in Congenital Heart Disease: from Fetus to Adult

Ageliki A. Karatza*

Department of Paediatrics, University of Patras Medical School, Patras, Greece

***Corresponding Author:** Ageliki A Karatza, Senior Lecturer in Paediatrics-Paediatric Cardiology, Department of Paediatrics, General University Hospital of Patras, Greece.

Received: May 17, 2017; **Published:** May 29, 2017

Keywords: *Congenital heart disease; Adult congenital heart disease; Antenatal diagnosis; Bioethics*

Abbreviations: CHD: Congenital Heart Disease; ACHD: Adult Congenital Heart Disease

Volume 1 Issue 1 May 2017

Introduction

Cardiovascular malformations represent the most prevalent birth defects in the human, having a variable reported incidence of 0.4-1.4% in live-born infants [1]. Major congenital heart disease (CHD) has life threatening complications, results in lifelong morbidity and represents nearly half of all infant deaths attributed to birth defects [2]. However, currently over 90% of children with serious CHD who have access to surgery survive to adulthood. No other cardiovascular intervention has achieved that kind of 20-year survival. As a result, there are currently 1.3 million adult patients with CHD, who now outnumber children with cardiovascular malformations [3].

Patients with operated CHD require life-long observation and specialized care. Even if the quality of the repair can be considered optimal, human intervention can are never imitate nature. Furthermore, some repairs establish unique physiologies that often manifest with serious complications later in life [4]. Long-term complications include atrial arrhythmias, pulmonary hypertension, and a repeated need for surgery, which result in significant increases in health services utilization during childhood, transition years, adulthood, and in the geriatric age group [3].

As these patients age and their population progressively enlarges, a number of ethical issues and social policy challenges arise. Unfortunately, there is scant empirical research examining either classical ethical issues in the CHD population or the unique ethical and policy challenges pertaining to CHD. Little emphasis has been given on the ethical aspects of the clinical guidelines and policy statements involving this population [5]. The first specialized meeting that brought together experts from the fields of ethics and CHD took place at the University of Pennsylvania in Philadelphia 2012 [6].

Ethical issues in prenatally diagnosed congenital heart disease

Prenatal diagnostic ultrasound is widely performed in the developed countries. Parents attending a scan expect to be told that their baby is normal. When a possible anomaly is identified and is subsequently confirmed, counseling is complex and requires much sensitivity. Since it is now possible to detect fetal cardiac anomalies prenatally, Paediatric cardiologists have to address the ethical issues involved in these clinical situations. Controversies arise as to how they will counsel a pregnant woman, how directive they will be about the preferable option and what they will do if the family's choice is not the one that optimizes fetal outcome [6]. The counseling process may take place in a stepwise fashion, sometimes over several medical sessions, and may need to be revisited, due to the emotional discomfort associated with the situation and the complexity of the information to be transferred.

The first stage is providing accurate information about the diagnosis and prognosis of the cardiac abnormality, in a manner and at a time that the expectant parents are able to understand [7]. Fetal anomalies necessitate that physicians provide the parent adequate information and credible counseling, an issue that is extremely relevant in the case of antenatally diagnosed congenital heart disease.

The next stage is to identify and present the options available to the family [7]. There are three main options: to continue with the pregnancy, to terminate the pregnancy (if legally permitted), or to consider prenatal intervention (if it is available for the condition). If the option to continue with the pregnancy is chosen, there will be further decisions to make such as where the infant is going to be delivered, the probable need for in utero transfer, and the mode of delivery. The parents should be aware of these implications, not in an attempt to change their decision, but to be informed and prepared them for the process.

The most ethically controversial step is discussing the options with the family [7]. There are different views about whether it is appropriate to recommend or favour a particular option, rather than being as neutral as possible. Another debatable issue is who is the more appropriate person to perform the counseling. What parents need most from the Paediatric cardiologist is the best available understanding of what their child's life would be like, what sort of interventions would be needed and the risks of these to the child in the local setting.

If intrauterine interventions are available for the current condition and local settings, the pregnant woman is in the ethical role of making decisions for the health of her future child, but she is also making decisions about her own health. Another challenging issue is dealing with a twin pregnancy where only one fetus has a serious cardiac anomaly [8]. The management depends on the wishes, values and preferences of the mother/parents once provided with detailed and accurate information about the condition of the twins. Any management plan incorporates risks to the unaffected fetus too, an issue that compounds the ethical considerations further and becomes more significant and complicated in monochorionic twinning.

"Palliative care" implies management that focuses on supportive, comfort measures when cure is not possible. The objective of palliative care is to improve the quality of life by lessening disease intensity and discomfort, while the condition runs its natural course [9]. In the era of antenatal diagnosis the number of patients presenting in shock or cyanosis after birth as the first indication of a cardiac anomaly is constantly getting uncommon. Therefore, depending upon the time of identification, families have the option of termination of pregnancy if the diagnosis is verified prior to 24 weeks of gestation. However, if diagnosis is made after 24 weeks of gestation or if termination of pregnancy is not desired for religious or cultural reasons, a third option that might be available (at least theoretically) to them would be to elect nonintervention at birth. The likelihood of a community sense of palliative care, as a reasonable choice is inversely proportionate to the capacity to successfully manage a particular disease and its prognosis. At some point, palliative care becomes inappropriate, once the threshold for true or perceived outcome achieves a satisfactory level [9].

In summary, conformance with the basic principles of "autonomy" and "beneficence" are the major ethical responsibilities of the Perinatal Cardiologist in pregnancies where there may be a serious fetal cardiac anomaly. The principle of "autonomy" creates a responsibility for the physician to help the pregnant woman make informed decisions based on her personal values and culture. On the other

hand, as it is possible for a fetus to survive in the extra-uterine environment independently (a state related to the degree of fetal maturity, which is further enhanced by the availability of medical technology) the ethical dilemma of whether the fetus is an independent subject that deserves "beneficience" becomes imminent. Therefore, a paramount ethical issue is achieving a balance between the pregnant woman's autonomy and the potential "beneficence" to the future child, as the fetus can reasonably be regarded as an independent subject [7].

Genetic Testing in Congenital Heart Disease: Ethical Considerations

Recent epidemiological studies have shown that about 30% of patients with CHD present a genetic syndrome or an association of cardiac and extracardiac anomalies [10]. Despite the impressive improvements in surgical repair of even the most challenging forms of CHD, patients featuring genetic syndromes may show an increased risk for death or major complications, requiring dedicated care in the postoperative period. These patients may not only represent a surgical challenge, but also suffer multiorgan impairments associated with their genetic condition [11].

As genetic testing becomes more and more inexpensive, concerns about privacy, use of genetic data for research, accuracy of testing, relationship of results to phenotypic expression, and uncertainty of prognosis continue to generate controversy [12]. If a genetic analysis is performed the results will be used, but the question is how, for what purposes and with what consequences [13]. Currently, our ability to acquire person specific DNA data far exceeds our knowledge of how to use this information in clinical practice effectively [12].

Inevitably, a number of crucial ethical issues continue to be a matter of debate:

1. Who, when, and why should we screen for genetic alterations in the congenital heart disease population?
2. What are the current risks and benefits of genetic testing?
3. How and by whom should the resulting information be transferred to patients and their families?

Genetic testing in this population is expected to have several benefits such as earlier detection of associated co-morbidities and timely interventions, as well as better understanding of potential or likely outcomes. Is also allows more accurate recurrence risk counseling, may offer the family an answer to the question ''why'' a certain medical condition is present to their child and might provide greater access to medical and social resources [12].

However, genetic testing of patients with CHD may encounter a number of significant risks as unreliable counseling could create an overly optimistic or pessimistic picture. The variability in phenotype and clinical course of many genetic syndromes may add further uncertainty to the prediction of outcome. Also, there is potential for insurability and employee discrimination or social stigmatization. Sometimes, identification of a genetic abnormality might prompt denial of access to specialized medical resources by the patient or the caregivers [12]. In the ensuing years, the debate over genetic testing in CHD patients will continue, although several notable changes have occurred with respect to medical knowledge, diagnostic techniques and legislative protection of the patients.

Ethics of Innovation in Surgery

Innovation in medicine involves the introduction of new methods, ideas, treatment modalities or devices to benefit an individual patient. Innovation in cardiothoracic surgery has resulted in tremendous advancements in intensive care techniques, novel strategies of myocardial protection, open heart surgery, as well as countless new operations, modified procedures, and new devices [13]. Innovation is a moral duty for physicians, however well-designed properly designed randomized, controlled trials provide the best evidence on the efficacy of novel health interventions [14,15].

Surgery is confounded by human factors such as skill and learning curves. Therefore, dilemmas as to whether all surgeons should be included in a clinical trial or only the more skilled ones often arise. If surgical innovation is considered as the progress in medical practice, it is governed by professional standards and standards of malpractice [16]. Responsible institutions require peer review of new operations and careful follow up of complications and outcomes. However, surgeons do not have to be re-accredited when they alter

practice or when they introduce new procedures. When there is very little knowledge, a significant amount of courage is required both from the part of the surgeon and the patient. As knowledge increases, the problem and its potential solution become more defined, which when applied to the care of the patient will require less courage in the plan of treatment [16].

Mechanical circulatory support as a bridge to heart transplantation

In patients with end-stage CHD disease, mechanical circulatory support may improve survival, quality of life, and serve as bridge to transplantation [17]. There are many ethical issues surrounding the use of mechanical circulatory support in patients with CHD including the use of prospective randomized controlled trials, proper oversight of new therapies, and transparency in reporting of results. Additionally, there are ethical considerations relevant to the greater society as these therapies are very expensive and the costs have to be covered by a resource-limited society. One of the important ethical issues surrounding ventricular assist devices, and any new therapy, is the process of evaluating whether this new therapy actually provides clinical benefit for the intended population. As newer devices become as a bridge to transplantation, inevitably increased demands are generated on an already scarce donor pool [17].

There are many unique considerations between adult and pediatric heart transplantation based on the inherent dependency of children, physiologic differences related to normal growth and development, and the heterogeneous etiology of heart disease and comorbidities [18]. Adult patients make autonomous decisions regarding their medical care. However, in pediatric medicine, autonomy as a principle is replaced by "beneficent paternalism", with parents making health care decisions for the child. In addition, unique to pediatric ethics, the wishes of a child's parents may be overruled by a clinician when the clinician believes that an alternative decision is in the best interest of the child. This moral obligation on the part of the pediatric practitioner exists due to the concept that the pediatric patient has a developing autonomy that must be respected.

However, the biggest challenge is the limited life span of the transplanted organ relative to the age of the child and the limited availability of donor organs [19]. The topic of re-transplant therefore arises and significant ethical issues regarding best allocation of a scarce resource become imminent. These issues, and likely new ones, are likely to become imminent as more patients with CHD are supported with ventricular assist devices, thus experiencing prolonged survival [18].

Ethical considerations and Clinical Trials in Pediatric patients with CHD

There is a strong need for well-designed clinical trials in CHD rather than extrapolating from different disease states in different age populations. The American Academy of Pediatrics supports this and believes that it is ''morally imperative to formally study drugs in children so that they can enjoy appropriate access to new and existing therapeutic agents" [20]. Research in children must adhere to the highest ethical standards at all times and this requires strict adherence to all regulatory requirements [21].

The research proposal must be scientifically sound and significant, including being appropriately powered. The research design needs to take into account racial, ethnic, gender, and socioeconomic characteristics of children and families. It is critically important that the investigators have the appropriate training to carry out the study. The study proposal needs to be directed by investigators in a state of scientific equipoise. The physicians must strive to prevent bias from affecting the design of the study, conduct of the study, or reporting of results [21]. They must ensure adequate disclosures of conflict of interest, maintain detailed medical records and comply with all regulatory, legal and ethical standards for research in children.

Regardless of the category of research, all pediatric research designs must take safeguards to protect participants. The recognition of children as a vulnerable population leads to a unique approach to risk-benefit assessment, external monitoring, and informed consent. Regulatory oversight of clinical trials in children is intended to enhance the safety and overall positive impact of those trials on children's health, particularly in the context of testing the safety and efficacy of new drugs, biologic agents, and devices that are to be used in children [22].

Ethics in Adult congenital heart Disease (ACHD)

Adult cardiologists are insufficiently trained in the care of patients with CHD, and, despite the recent establishment of training programs for adult congenital heart disease (ACHD), there is an inadequate supply of clinicians with expertise in the field. As a consequence, young adults and even older adults with complex CHD continue to be followed-up in the Paediatric Cardiology outpatient clinics. As these patients grow older, they may develop ''adult'' health problems such as coronary artery disease, and the quality of provided medical care may become suboptimal, as their management is fragmented between specialists [4].

Even worse, the young adult patient with CHD may not follow the instructions to make appointments with the ACHD team, and get lost in the transition and re-present in the acute and emergency Department of the local hospital with cardiac decompensation that could have been potentially avoided with routine surveillance and proactive care. In the era of ''patient centered medicine,'' transitions are impacted by psychosocial challenges, cultural adaptation, religion/spiritual needs, and intrafamilial dynamics. ''Shared care'' programs must address the concerns and needs of patients in these domains, as well [4].

The CHD population presents unique challenges in addressing advance care planning and palliative care. For patients and families who have benefitted from high-technology interventions, encouraging advance care planning, including discussion of limitations on further interventions or withdrawing therapies, can be challenging. An organized communication between adults with CHD and their cardiovascular clinician is particularly critical when discussing long-term planning, whether years in advance or near the end of life. Clinicians should acknowledge that death will happen at some point and provide realistic expectations for the patient's course and give honest answers to patients and their families [23].

The principle of "autonomy' is central to participatory or informed decision making, which engages the patient and incorporates their preferences for the end of life and aggressiveness of care. In participatory decision making, physicians and other clinicians guide decisions based upon what is medically possible and reasonable for the individual patient. Based upon the concepts of "beneficence" and "non-maleficence", we should strive to optimize care for patients at the end of their lives and include palliative care, as appropriate. The ultimate goal is to provide minimal distress and pain for patients, the family members and the care team [23].

References

1. Hoffman JIE and Kaplan S. "The incidence of congenital heart disease". *Journal of the American College of Cardiology* 39.12 (2002): 1890-1900.
2. Richmond S and Wren C. "Early diagnosis of congenital heart disease". *Seminars in Neonatology* 6.1 (2001): 27-35.
3. Marelli AJ., *et al*. "Lifetime Prevalence of Congenital Heart Disease in the General Population from 2000 to 2010". *Circulation* 130.9 (2014): 749-756.
4. Kirkpatrick JN., *et al*. "Ethics Priorities in Adult Congenital Heart Disease". *Progress in Cardiovascular Diseases* 55.3 (2012): 266-273.
5. Kirkpatrick JN and Kaufman B. "Why Should We Care About Ethical and Policy Challenges in Congenital Heart Disease?" *World Journal for Pediatric and Congenital Heart Surgery* 4.1 (2013): 7-9.
6. "Ethics of the Heart: Ethical and Policy Challenges in Adult and Pediatric Congenital Heart Disease".
7. Malhotra A., *et al.* "Ethical Issues in Fetal Management: A Cardiac Perspective". *International Journal of Pediatrics* 1 (2010): 1-6.
8. Malhotra A., et al. "Medical and ethical considerations in twin pregnancies discordant for serious cardiac disease". *Journal of Perinatology* 29.10 (2009): 662–667.
9. Rychik J. "What Does Palliative Care Mean in Prenatal Diagnosis of Congenital Heart Disease?" *World Journal for Pediatric and Congenital Heart Surgery* 4.1 (2013): 80-84.
10. Grech V and Gatt M. "Syndromes and malformations associated with congenital heart disease in a population-based study". *International Journal of Cardiology* 68.2 (1999): 151-156.

11. Formigari R., *et al*. "Genetic syndromes and congenital heart defects: how is surgical management affected?" *European Journal of Cardio-Thoracic Surgery* 35.4 (2009): 606-614.
12. Lin KY., *et al*. "Genetic Testing in Congenital Heart Disease: Ethical Considerations". *World Journal for Pediatric and Congenital Heart Surgery* 4.1 (2013): 53-57.
13. Philippidis A. "Panel Wrestles with WGS Ethics: Final Recommendations Will Encompass Technology Delivery and Informed Consent". *Genetic Engineering and Biotechnology News* 32.7 (2012): 6-8.
14. Altman DG., *et al*. "The revised CONSORT statement for reporting randomized trials: explanation and elaboration". *Annals of Internal Medicine* 134.8 (2001): 663-694.
15. Moher D., *et al*. "The CONSORT statement: revised recommendations for improving the quality of reports of parallel-group randomized trials". *BMC Medical Research Methodology* 1 (2001): 2.
16. Mavroudis C., *et al.* "Ethics of innovation in surgery for congenital cardiac diseases". *Cardiology in the Young* 19.2: 100-105.
17. Rossano JW., *et al*. "Ethical Considerations Related to the Use of Mechanical Support in Congenital Heart Disease". *World Journal for Pediatric and Congenital Heart Surgery* 4.1 (2013): 70-74.
18. Kaufman BD and Jessup M. "Adult and Pediatric Perspectives on Heart Retransplant". *World Journal for Pediatric and Congenital Heart Surgery* 4.1 (2013): 75-79.
19. Kirk R., *et al*. "The Registry of the International Society for Heart and Lung Transplantation: Thirteenth official pediatric heart transplantation report--2010". *The Journal of Heart and Lung Transplantation* 29.10 (2010): 1119-1128.
20. Shaddy RE and Denne SC. "Clinical report--guidelines for the ethical conduct of studies to evaluate drugs in pediatric populations". *Pediatrics* 125.4 (2010): 850-860.
21. Shaddy RE. "Ethics and Clinical Trials in Congenital Heart Disease". *World Journal for Pediatric and Congenital Heart Surgery* 4.1 (2013): 50-52.
22. Flotte TR., *et al*. "Recent developments in the protection of pediatric research subjects". *The Journal of Pediatrics* 149.3 (2006): 285-286.
23. Kovacs AH., *et al.* "Advance Care Planning and End-of-Life Management of Adult Patients with Congenital Heart Disease". *World Journal for Pediatric and Congenital Heart Surgery* 4.1 (2013): 62-69.