Review Article

Critical cardiac care in children: looking backward and looking forward

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Abstract: The growth of Pediatric Cardiovascular Intensive Care as a subspecialty has been incredible. Outcomes have improved, care delivery has matured, and research has made advances. Within this review, we take the opportunity to examine the subspecialty's past accomplishments with pride, take stock in its current state, and look forward with excitement to its future. While outcomes in general have improved dramatically, we must always be aware of the outcomes that matter to families and patients. Additionally, we must constantly ask ourselves to improve. Research into neuroprotection and individual therapeutic strategies based in genomic medicine provide the next opportunity for the subspecialty to improve.

Keywords: Pediatric; cardiac; congenital heart disease (CHD); outcomes

Submitted Jun 16, 2016. Accepted for publication Jun 24, 2016.
doi: 10.21037/tp.2016.06.07
View this article at: http://dx.doi.org/10.21037/tp.2016.06.07

Introduction

Pediatric Cardiovascular Intensive Care has become increasingly organized as a subspecialty over the past two decades. The pediatric cardiac intensivist plays a central role in the critical care of these patients, as well as continuous quality improvement and family centered care. This growth of the subspecialty comes in response to the explosion of knowledge and research in the patient with critical cardiac disease, the increasing complexity of cardiac lesions and procedures to treat them, and the growing numbers of patients of a younger age requiring cardiac intensive care. Indeed an international subspecialty society, the Pediatric Cardiac Intensive Care Society, was organized in 2003 to address the issues facing practitioners.

Within this review, we take the opportunity to examine the subspecialty's past accomplishments with pride, take stock in its current state, and look forward with excitement to its future. Additionally, it gives the opportunity to applaud those who attempt to innovate in order to radically improve the future care of these children.

Looking backward

It is clear that we have had rapid advancement in all outcome measures (1). However, the danger of always looking backward is that we are subject to either positive or negative revisionist history. Additionally, hindsight is always 20/20. The reality is, as with all history, truth is found somewhere in the middle ground between the superlative and stupidity. We were never as good, nor as bad, as we think.

An interesting conceptual framework that is important in medicine is that throughout the history of care, at the time and in the present, we were convinced that we were doing the right thing for our patients. However, many of these truths have subsequently proved to be false. In the modern world facts change all of the time, according to Samuel Arbesman, author of *The Half-Life of Facts*: *Why Everything We Know Has an Expiration Date* (2). Since scientific knowledge is still growing by a factor of ten every 50 years, it should not be surprising that many of the facts people learned in school and universities have been overturned and are now out of date. But at what rate do former facts disappear? Applying the concept of half-life to facts, Arbesman cites research that looked into the decay in the truth of clinical knowledge about cirrhosis and hepatitis. "*The half-life of truth was 45 years*," reported the researchers.

An example of the changing "truth" occurred in relation to George Washington, the former President and senior leader of our nascent country. On December 12, 1799,

Washington suffered from an upper respiratory infection (3). His physicians applied a painful "blister of cantharides", better known as "Spanish fly", to Washington's throat to cause "counter-irritation". They justified the removal of more than 80 ounces of his blood (2.365 liters or 40 percent of his total blood volume) over a 12-hour period in order to reduce the massive inflammation of his windpipe and constrict the blood vessels in the region.

Of course, this is seen as ridiculous in today's scientific understanding. However, as stated, in their present it was the most justifiable approach. So with this as background, what are some of the "truths" in the care of critically ill children with cardiac disease that will be questioned in the future. I propose discussion pertaining to three present day "truths". First, we think surgeons actually matter. Second, we think doctors actually matter. Finally, we think we know which outcomes matter.

We examined the influence of surgical volume on outcome in a recent investigation of the Norwood procedure (4). Lower mortality following the Norwood procedure was associated with high institutional volume. However, lower mortality was not associated with the number of cases performed by a surgeon. We concluded that a well-experienced surgeon was necessary but insufficient to truly impact positive outcomes. The impact of the institution, the team, had a greater influence on outcomes.

Ultimately, cardiovascular critical care is a team sport. Every participant has a role in the care of each child. Everyone from physicians, to nurses, to therapists, to family members, all influence the success of complex care. However, there is a struggle within this team concept. The team is important, but ultimately the individual is accountable for their performance. As stated by former coach Phil Jackson, "*The strength of the team is each individual member...the strength of each member is the team*". It is incumbent on the specialty as a whole to develop care models that enhance teamwork while maintaining a culture of individual skill, pride and accountability.

Another team member that is viewed as important in today's truth construct is the physician. I would contend that doctors are not as important as we believe ourselves to be in today's cardiovascular intensive care unit. The most important member of today's unit team is the bedside nurse.

While we continually rely on technology in the form of monitors, diagnostic imaging, and laboratory surveillance, all data gained from monitors must be integrated with the information gained by physical exam. An experienced clinician must accomplish this integration. While technology can serve to aid in the care of the patient, nothing can replace the experience of a clinician. Bernard Lown, writing in *Scientific American*, outlined such a balance over 40 years ago. "*Neither monitors nor the most complicated electronic gear makes a coronary care unit. The fundamental ingredient is a properly indoctrinated nursing staff. The reason for this is obvious. The nurse is usually the only trained medical professional at the bedside during important clinical events. The time for effective action is brief and does not usually allow delay for the arrival of a physician. The nurse is trained in the recognition of arrhythmias and is delegated the authority for enacting the entire repertory of lifesaving techniques In fact, many well-functioning coronary care units have been successful because of the elite spirit and competence of the nursing staff.*" (5).

As was apparent in the infancy of cardiac critical care, the presence at the bedside by experienced clinicians was paramount to success. However, this paradigm is currently under attack. We are forced to limit the experience gained by trainees and bedside nurses. While we profess the desire to avoid monitors acting as a replacement to experienced clinicians, we are forced to re-examine their utility when faced with shifts covered by residents and fellows who are restricted by work hours, and young nurses who have just graduated nursing school. This represents a new challenge to the continued growth and success of our care delivery models.

Finally, we think we know the outcomes that matter. Boneva *et al.* (1) reviewed population-based mortality data of congenital heart disease (CHD) from 1979–1997, from the Center for Disease Control and Prevention (CDC). Overall mortality decreased 39%. Yet there was a smaller decline in HLHS—7.5%, and TOF—10%. The decrease for TGV was 40.6% in infants <1 year and 74.4% in children 1–4 years of age. In fact, our center has reported overall outcomes of surgical procedures improved to <1% mortality. While individual lesions, risk categories, and co-morbid conditions impact this low mortality leading to variations within risk subcategories, the improvement is obvious. We are not alone in this staggering improvement. It is safe to say that, as a community, we have moved CHD from an expected mortality to an expected survival. However, this creates risks, opportunities, and consequences, not the least of which is a loss of perspective of meaningful outcome.

In 1986, Lillehei *et al.* reported long term follow up on his first operations conducted from 1954 to 1960 (6). Of course, he was proud of an actuarial survival at 30 years of 77%. However, he also went on to highlight other outcomes. In his

cohort of patients, 32% completed college, ten completed graduate school, 40 patients had children with 93% of those being live births, and 7.3% with cardiac defects. He understood that these are the outcomes that truly matter. This pre-dates the latest attention to neurodevelopmental outcomes.

Ever since Alfred Blalock reported how successful the original interventions were at improving quality of life (7), we, as a community, thought we were great. However, that greatness was short lived. Ignorance, it seems, truly was bliss. As these children aged, we realized the impact of CHD, surgical interventions, cardiopulmonary bypass (CPB), and medications on the neurodevelopmental outcomes of these children (8). Our historical success has allowed us to realize that the road to an adult survivor of CHD is one that is far from linear. It is now incumbent on our entire field to cooperate, coordinate, and collaborate to determine how best to protect the neurologic status of our patients, and allow them to become the type of adult survivor we hope for when we first meet with families.

Looking forward

This easily transitions to looking forward in our subspecialty. Where do we go from here? How do we improve? The path to improvement in care involves education, research, and innovation. It is through the combined work of the committees and the Board of Directors of the PCICS that will soon yield training pathways for physicians and nurses seeking additional experience in cardiac critical care, international quality improvement initiatives, online journal clubs, and a research structure that will provide robust collaboration and mentorship opportunities. Innovation requires pushing boundaries, changing perspective on current problems, and taking risks. Three areas that have promise to do just that involve protection during CPB, data management, and the promise of individualized medicine.

In a recent study, we evaluated the impact of delivery of the gas nitric oxide (gNO) to the membrane oxygenator of the CPB circuit on postoperative outcome measures in children undergoing cardiac surgery for CHD (9). Children who received gNO during CPB had an improved postoperative course, as demonstrated by significantly reduced myocardial injury and shortened duration of mechanical ventilation and length of stay in the pediatric CICU. This has been reproduced by colleagues in Australia (10). Our premise is that NO added to the circuit has effect distal to the entry site. This is a novel concept and one that fits the requirement of pushing boundaries for innovation. It is possible that through this type of novel, innovative application of existing drugs or therapies, we may impact outcomes in ways not previously realized.

Innovation is also necessary to adequately capture and interpret the ever-expanding wealth of data generated by each individual patient or event within a critical care hospitalization. Ultimately, data equals power. Data gives us the power to do the right thing well, at the right time and with the minimum of resources. Do it well, once, and with no complications.

For example, monitoring patients allows us to gauge the effectiveness of our efforts. Our goal is to monitor, and then intervene, in order to avoid progression to a decompensated shock state. It is the cornerstone of modern critical care medicine that intervening prior to the development of end-organ dysfunction or damage yields improved outcomes for the patients in our care. While monitoring can guide intervention, one effect of this approach is the generation of increasing volumes of data. As an intensivist, we must manage an enormous amount of information each moment we care for patients. These data must ultimately guide interventions. Yet with the growing volume of data, how do we know what information is meaningful? How do we separate the wheat from the chafe? This is the role of effective monitoring and effective data management in a modern ICU setting.

The second challenge for innovation comes in the integration of the overwhelming amount of data presented in a modern pediatric cardiac intensive care setting. We not only manage patients, we manage data. We need to develop the means to adequately monitor trends and pick up a signal when one is present. Monitoring in the pediatric cardiac intensive care environment should be an intuitive and analytic process. As noted above, there are numerous monitoring modalities available, both physiologic and laboratory based. The clinician at the bedside needs to be able to integrate this information to track the trajectory of the patient, and decide on interventions when necessary. Further, we do not know the impact of specific monitoring on patient recovery and outcome, on cost effectiveness and on the longer term quality of life; we assume we are monitoring the right predictors of outcome and that the target ranges are correct as well.

We have assumed that more is better although there are clear problems with fixation on specific abnormal results that deflect critical decisions. We now work in very complex environments. There is a huge amount of information

coming to the clinicians from physiologic and laboratory data, yet we do not collect, store and analyze this data in real time. In addition, there are multiple distractions at the bedside with continual interruptions to workflow. In short, we have not leveraged information systems to our benefit, and have not leveraged our common knowledge within the field and between institutions to standardize care and resource utilization. We need to leverage monitoring data to move away from the traditional "chain-of-event" analysis following adverse outcomes, which focuses primarily on patient characteristics and human error, and also move away from a "failure to rescue" analysis which focuses on unit-based team structure and function. Rather, we should focus on "failure to predict" an evolving clinical picture, which really evaluates systems characteristics and data integration. We need to understand how we function as a system and leverage the information systems to support our workflow.

In addition to information systems and data management, there has been an explosion of genetic data and power in the past two decades. We now have the ability, through whole genome informatics, to analyze the information found in literally thousands of genes within minutes. Data from investigators such as Hector Wong and Perren Cobb (11,12), to name but two, indicate that blood transcriptional and proteomic profiles can distinguish between host responses to different types of injuries in different age groups. They are demonstrating that information at the *genome* (DNA) level provides information about predisposition to a given outcome, while data at the transcriptome (RNA) (13) and proteome (protein) levels can be harnessed to make diagnoses, and finally gauge the response to therapy (prognoses). The promise of this line of investigation is that these patterns of change in gene and protein expression, in effect, become new, genomic "vital signs" (14). Additionally, we now have the computational power to not only analyze these data at a single point in the time course of a patient, but also across the time spectrum of the entire disease and healing trajectory (15,16). Through these discoveries, we finally have the potential for truly personalized diagnosis and intervention. Within these lines of investigation lies the opportunity for providing the right care to the right person at the exact right time. What if we applied this approach and this technology to other outcome questions such as sedation and analgesia postoperatively, nutrition, and neurodevelopmental outcomes?

Conclusions

There have been incredible advances in the care of children with cardiac disease. We should all take pause and recognize the advancements that have been made. I would contend that there are very few areas of medicine that have achieved the same degree of success over the past 50 years. However, it is now incumbent on each of us in the field to build upon these advances so that the next generation of practitioners will be just as proud to look back on our latest accomplishments.

Acknowledgements

None.

Footnote

Conflicts of Interest: The author has no conflicts of interest to declare.

References

1. Boneva RS, Botto LD, Moore CA, et al. Mortality associated with congenital heart defects in the United States: trends and racial disparities, 1979-1997. Circulation 2001;103:2376-81.
2. Arbesman S. The half-life of facts: why everything we know has an expiration date. New York: Current, 2012.
3. Witt CB Jr. The health and controversial death of George Washington. Ear Nose Throat J 2001;80:102-5.
4. Checchia PA, McCollegan J, Daher N, et al. The effect of surgical case volume on outcome after the Norwood procedure. J Thorac Cardiovasc Surg 2005;129:754-9.
5. Lown B. Intensive heart care. Sci Am 1968;219:19-27.
6. Lillehei CW, Varco RL, Cohen M, et al. The first open heart corrections of tetralogy of Fallot. A 26-31 year follow-up of 106 patients. Ann Surg 1986;204:490-502.
7. Taussig H, Blalock A. Surgery of congenital heart disease. Br Med J 1947;2:462.
8. Marino BS. New concepts in predicting, evaluating, and managing neurodevelopmental outcomes in children with congenital heart disease. Curr Opin Pediatr 2013;25:574-84.
9. Checchia PA, Bronicki RA, Muenzer JT, et al. Nitric oxide delivery during cardiopulmonary bypass reduces postoperative morbidity in children--a randomized trial. J Thorac Cardiovasc Surg 2013;146:530-6.
10. James CS, Horton S, Brizard C, et al. Abstract 14827: Nitric Oxide During Cardiopulmonary Bypass Improves Clinical Outcome: A Blinded, Randomized Controlled Trial. Circulation 2015;132:A14827.

11. Chung TP, Laramie JM, Province M, et al. Functional genomics of critical illness and injury. Crit Care Med 2002;30:S51-7.
12. Wong HR. Genetics and genomics in pediatric septic shock. Crit Care Med 2012;40:1618-26.
13. McDunn JE, Muenzer JT, Rachdi L, et al. Peptide-mediated activation of Akt and extracellular regulated kinase signaling prevents lymphocyte apoptosis. FASEB J 2008;22:561-8.
14. Cobb JP, Brownstein BH, Watson MA, et al. Injury in the era of genomics. Shock 2001;15:165-70.
15. Wong HR, Cvijanovich N, Allen GL, et al. Genomic expression profiling across the pediatric systemic inflammatory response syndrome, sepsis, and septic shock spectrum. Crit Care Med 2009;37:1558-66.
16. McDunn JE, Husain KD, Polpitiya AD, et al. Plasticity of the systemic inflammatory response to acute infection during critical illness: development of the riboleukogram. PLoS One 2008;3:e1564.

Cite this article as: Checchia PA. Critical cardiac care in children: looking backward and looking forward. Transl Pediatr 2016;5(3):160-164. doi: 10.21037/tp.2016.06.07