Practice, Commentary, and Opinion

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Creating a Lesion-Specific "Roadmap" for Ambulatory Care Following Surgery for Complex Congenital Cardiac Disease

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Abstract

Over the past 20 years, the successes of neonatal and infant surgery have resulted in dramatically changed demographics in ambulatory cardiology. These school age children and young adults have complex, and in some cases, previously unexpected cardiac and non-cardiac consequences of their surgical and/or transcatheter procedures. There is a growing need for additional cardiac and non-cardiac subspecialists, and coordination of care may be quite challenging. In contrast to hospital-based care, where inpatient care protocols are common, and perioperative expectations are more or less predictable for most children, ambulatory cardiologists have evolved strategies of care more or less independently, based on their education, training, experience, and individual styles, resulting in highly variable follow-up strategies. We have proposed a combination proactive-reactive collaborative model with a patient's primary cardiologist, primary care provider and subspecialists, along with the patient and their family. The goal is to help standardize data collection in the ambulatory setting, reduce patient and family anxiety, increase health literacy, measure and address the non-cardiac consequences of complex cardiac disease, and aid in the transition to self-care as an adult.

Background

In 1980, Fyler and colleagues from the New England Regional Infant Cardiac Program coined the term "critical congenital heart disease" (cCHD); defined as CHD severe enough to require surgery or cardiac catheterization before 1 year of age.[1] At that time, the prognosis for many neonates with cCHD was grim, as

- stabilization with prostaglandin E_1 had just been approved by the Food and Drug Administration in the United States of America,
- two dimensional echocardiography was in its infancy, and
- the fields of cardiac intensive care, perfusion, anesthesia and neonatal surgery were less developed.

Mortality rates were high following surgery in the first month of age, and the longer-term outcomes were purely speculative. Those with cCHD surviving the neonatal period without surgery to reach infancy frequently received palliative procedures prior to definitive correction at a later date; typical examples of such patients include those with

- transposition of the great arteries with a ventricular septal defect,
- common arterial trunk,
- tetralogy of Fallot, and
- large ventricular septal defects or atrioventricular septal defects

Over the past 20 years, the successes of neonatal and infant surgery, including "corrective" procedures for those with two ventricles and "palliative" procedures for babies born with a functionally univentricular heart, have resulted in dramatically changed demographics in ambulatory cardiology. These school age children and young adults have complex, and in some cases, previously unexpected cardiac consequences of their surgical and/or transcatheter procedures[2-29] (Table 1). Some of these consequences seem to affect many of these patients to some degree, such as:

- diminished exercise performance,
- chronotropic impairment, and
- a risk of obesity and sedentary lifestyle.

Meanwhile, some of these consequences are lesion-specific, such as:

- arrhythmia,
- valvar regurgitation, and
- asymmetric pulmonary blood flow.

In addition, there is a growing recognition of additional non-cardiac consequences, including:

- challenges with school performance,
- social disintegration,
- anxiety and depression,
- restrictive lung disease,

- reactive airways disease,
- sensori-neural hearing loss,
- reduced health-related quality of life,
- significant medication burden,
- post-traumatic stress (for the patient and the family),
- anti-social behavior, and
- challenges with employment and health care insurance.

For those with a functionally univentricular heart, additional burdens may include:

- delayed puberty and short stature,
- thrombosis,
- renal dysfunction,
- protein losing enteropathy,
- cirrhosis, and
- plastic bronchitis.

As these interdisciplinary challenges have become more apparent, ambulatory cardiology has become increasingly complicated for these patients, and now consists of much more than simply

- physical examination including auscultation,
- an electrocardiogram,
- the evaluation of an echocardiogram, and
- discussion of recommendations.

There is a growing need for additional cardiac and non-cardiac subspecialists, and coordination of care may be quite challenging. Unfortunately, routine office visits are frequently time-constrained, particularly for those with the most complex disease and/or multiple cardiac and non-cardiac consequences. In fact, time may not allow for all information and counseling to be given. Some questions of a patient or family may go unaddressed.

Traditionally, ambulatory care focused on health maintenance and management of acute changes in health, but there is now a growing need for chronic interdisciplinary care, sometimes termed the "medical home". In 2004, Palfrey et al. introduced the concept of the pediatric alliance for coordinated care. The study consisted of a pediatric nurse practitioner that was assigned as the case manager of children with special healthcare needs; most with severe chronic illness, and >5 conditions. In one study,[30] after 2 years of utilization of the "medical home", evaluations by the family (n=117) of the "medical home" were obtained and revealed marked improvement in patient satisfaction scores in areas including:

- a decrease in emergency room visits throughout the year,
- a decrease in sick days taken from work,
- improved accessibility to resources,
- earlier intervention when the child was acutely ill, and
- enhanced communication with the medical team.

Current approach to outpatient follow-up

As the focus of outcome studies shifts to the long-term, outcomes in the ambulatory setting play an increasingly important role. The outcomes measured in childhood, adolescence and young adulthood not only describe the patient cohort, they serve to inform and modify earlier care strategies. However, in contrast to hospital-based care, where inpatient care protocols are common, and perioperative expectations are more or less predictable for most children, ambulatory cardiologists have evolved strategies of care more or less independently, based on their education, training, experience, and individual styles. Systematic collection of data may occur for specific, targeted research proposals, typically utilizing a chart review or cross-sectional study design, with the limitations and biases inherent in those approaches. The care local cardiologists provide is likely to vary because there have been few standards for these children as there has been little systematically collected data to inform their care. Instead, clinicians may be influenced by multiple factors leading to variability and potential inefficiencies in follow-up strategies, including the potential for over-testing; these factors, including recall bias, the struggles of "that last difficult case", concern of "missing something", or worse, the concept of "I had a patient once who…". In some developed countries, there may be

financial incentives to perform more diagnostic testing, while in others, there may be financial incentives to do just the opposite.

To illustrate the variability in outpatient practices following surgery for cCHD, between 10 October 2006 and 4 November 2006 we used the internet to conduct a survey of ambulatory pediatric cardiologists to determine strategies used for follow-up after the Fontan operation, arterial switch operation and repair of tetralogy of Fallot (previously unpublished data, presented at the Scientific Sessions of the American Heart Association in 2006). A previously piloted questionnaire consisting of 43-items was distributed via an internet listserve (PediHeart) and known email addresses. Demographics of the survey respondents (n=434) are shown in Table 2, and results in Figure 1A-G. While this survey was conducted approximately 10 years ago, the results are likely to be similar if conducted today, and show significant practitioner-based variability in diagnostic testing. Preliminary analysis of the data suggested that more "senior" cardiologists performed diagnostic testing less frequently, but due to the subjective nature of the responses, and lack of subsequent validation, formal statistical analysis was not undertaken.

This "case by case" approach in the ambulatory setting, however, has important drawbacks:

- Unless the cohort is well defined and within one health care network or academic setting, the results of ambulatory testing may not provide feedback to the surgical and inpatient team, limiting or delaying modifications of technique to address problems.
- There are often too few patients at any one center or practice to inform local care guidelines. Frequently, follow-up of a surgical cohort is spread among many different clinicians, frequently in multiple locations.
- The cost-benefit analyses of follow-up testing are difficult if not impossible to undertake.

Indeed, in their landmark publication "Crossing the Quality Chasm", the Institute of Medicine stated "patients should receive care based on the best available scientific knowledge. Care should not vary illogically from clinician to clinician or from place to place."[31] The report further states that clinicians and institutions should actively collaborate and communicate to ensure an appropriate exchange of information and coordination of care.

Recently, landmark work by the team at Boston Children's Hospital has resulted in the development of standardized clinical assessment and management plans: "SCAMPs."[32-37] This initiative was produced as a means to reduce variation in practice and promote standardization, with the ability to individualize the care provided and thereby provide a means for ongoing modification of the plan. This breakthrough approach to care was first initiated for the care of children with cCHD due to variation in cardiology practices. Similar to this approach, the goal of a "roadmap" approach is to decrease the variability in medical practice, while allowing for individual variation in practice and collection of data. In most non-academic settings, collection of data and evaluation of strategies of surgical care may face challenges when multiple cardiologists throughout the community provide postoperative care, a care model that centers on individual providers rather than centers. Developing a roadmap for children born with cCHD is meant to be complimentary to a "SCAMP" approach, and is community-based as well as patient-based; such a roadmap will provide

- pre-set parental expectations for the type and timing of routine surveillance,
- an understanding of the expected consequences of surgery for cCHD, and
- a framework for screening that will be necessary over the lifetime of the patient.

In 2006, following meetings to determine consensus at The Children's Hospital of Philadelphia, establishing follow-up guidelines for patients with cCHD was attempted,[38] but the proposal did not meet with sustained success. As has been previously described by Cabana and colleagues, the inertia of the individual practice was (and is) hard to overcome.[36, 37, 39, 40] A number of additional barriers became apparent, as in many guidelines, including:

- provider lack of agreement,
- resistance or lack of knowledge of the guidelines,
- an inadequate database for tracking outcomes, and
- at the time, an electronic health record with only rudimentary capabilities to identify patients who would best be served by these guidelines and the appropriate intervals for follow-up and testing.

In addition, the plan may have been too ambitious, with yearly or biannual recommendations of diagnostic testing. Many of these factors have changed in the last decade, as bioinformatics and the electronic health record have matured.

Lost in all of this variability of management have been the patient and family. Many go from year to year, hoping nothing will "come up" during the annual or biannual visit, reassured by the "see you next year" and "everything looks great" sometimes paternalistic style of medicine. Results of tests may not be communicated in a manner that is understood. Barriers of language may exist. Health literacy, a key component to improved health-related quality of life and improved medical compliance, is quite variable across patients, and may be related to socioeconomic status as well as the interest and experience of the provider. The anxiety of the annual check-up can be severe at times, leading to incomplete "hearing" and processing of the information presented. Casually stating that a new test "might be helpful" can be received in a variety of ways, but in our experience, may lead to anxiety, uncertainty, and worry, and some families re-live the trauma of the initial diagnosis and uncertainty of the future. Transition programs to self-care are sparse or non-existent in some areas,[18] although on-line resources are recently available.[41] Consultants for the non-cardiac consequences are frequently at a disadvantage. Often, these consultants may never have seen a child or young adult with the particular cCHD, and may not see the relationship of the problem in "their organ system" to the underlying cCHD. This is particularly problematic in patients with neurodevelopmental challenges, and in those with a functionally univentricular heart and multisystem consequences.

Strategy

We have proposed a combination proactive-reactive collaborative model with a patient's primary cardiologist, primary care provider, and family in order to address:

- the complexities of cCHD consequences (or "complications" as they are sometimes referred to),
- the variability of the timing and severity of presentation, and
- the need for non-cardiac specialists.

To illustrate the combination proactive-reactive model, we frequently use the analogy of planned maintenance in the automotive industry. When we purchase an automobile, we are given a suggested maintenance schedule, a plan of surveillance for potential mechanical issues that are suggested at predictable intervals. This represents a "proactive" strategy, usually based on time or miles or kilometers driven, and not because the owner identifies a problem. Not each "checkup" is comprehensive, but at important epochs, for example, at 100,000 kilometers, a very comprehensive evaluation is done, in contrast to a less complete evaluation done every 10,000 kilometers. If the owner senses something unusual in the drive of the automobile, or the wipers need changing, a "reactive" strategy is always available, and is analogous to an unplanned visit to the cardiologist. Many families have embraced this approach, with "major" check-ups occurring infrequently but coordinated by the surgical center ("dealership"), and less comprehensive and/or unplanned visits occurring locally ("the local expert mechanic"). This strategy results in a number of major benefits:

- from the time of the surgical procedure, the family knows what to expect moving forward, when certain tests will be ordered, what the purpose of the test is, and what will be done with the results,
- the primary care provider and primary cardiologist direct these evaluations locally whenever possible, communicating the results with the surgical center, and
- over time, consistency in follow-up creates a database of "expected" findings and consequences for each particular cCHD.

The Roadmap: Standardized Testing and Routine Surveillance ("STARS") for cCHD

A good deal of thought was put into the frequency of surveillance testing. These recommendations are likely to be different in different scenarios, including:

- different models of care
- different socioeconomic situations,
- rural versus urban settings,
- free-market health care versus single-payer national health coverage, and

- many more.

Nevertheless, it seems reasonable from a developmental perspective, fiscal perspective, as well as extensive review of the literature and discussion with families, that five planned visits between surgery and transition to self-care as an adult is a good starting point (see Figure 2). Importantly, these recommended comprehensive evaluations include non-cardiac evaluations and recommendations. We advocate comprehensive evaluations at the following five times:

- at the first birthday;
- upon entering elementary education [~5-6 years of age];
- between elementary and middle school [~10-11 years of age];
- during transition between middle school and high school [~14-15 years of age], and
- upon transfer to adult care [~18-21] years of age.

These times are just starting points for discussion, but setting up a lifelong plan for patients and their families has multiple advantages; such a "Roadmap":

- manages expectations and anxiety,
- leads to in-depth conversations about the cCHD and current status of the repair, improving health literacy,
- emphasizes the need for life-long care, particularly if introduced early in follow-up,
- emphasizes and evaluates the safety and importance of exercise,[42,43] and primary prevention of atherosclerotic cardiovascular disease,[44,45]
- leads to simultaneous investigations of other organ systems, particularly neurodevelopment, as recently recommended by the American Heart Association and American Academy of Pediatrics,[46] and
- may aid in transition and self reliance.[18,47]

Following the comprehensive "100,000 kilometer checkup", a full summary of the results, including interpretation, is reviewed with the patient's primary cardiologist, primary care provider, and family, with all questions addressed as best as possible.

Examples of the Roadmap to be implemented at The Heart Program at Nicklaus Children's Hospital are included in the appendix. Note that for each lesion, there are specific tests for known consequences specific to the repair, as well as more general screens for consequences such as:

- exercise performance,
- level of activity,
- arrhythmia,
- obesity,
- neurodevelopment, and
- general health status.

It is also important to recognize and emphasize the importance of the first year of life following surgery for cCHD. Some of these patients remain physiologically fragile following palliative or corrective procedures. Many, particularly those requiring intervention in the first weeks of life, have a higher incidence of challenges with feeding, nutrition, heart failure, developmental delay, medication burden and a heightened level of family stress and anxiety. For neonates, with a functionally univentricular heart, many programs have instituted an intensified level of ambulatory surveillance and support with "interstage monitoring programs", but we would argue that the same level of support, and interdisciplinary involvement should be extended to infants with biventricular repairs, particularly if there are multi-system challenges as mentioned above. Setting up the framework and expectations for interdisciplinary follow-up and family support should start immediately after the surgical repair, and continue throughout childhood. Critical in this process is a structured "hand-off" from the surgical center to an interdisciplinary team, lead by the child's primary cardiologist and primary care provider.

One may argue that it is over ambitious, or perhaps not indicated, for the cardiology program where surgery was performed to provide the medical home for these patients. Some may feel that it is not the responsibility of the cardiologist to evaluate non-cardiac domains, including:

- neurodevelopment,

- growth failure,
- sedentary behaviors, and
- lack of separation from parents.

We disagree. While it is certainly not the responsibility of the cardiologist at the surgical center to provide all of these services, we feel it is the responsibility of that child's cardiologist to coordinate the care team, and know what colleagues need to collaborate in order to provide the medical home, along with the child's primary care physician, primary cardiologist, and family. The cardiology team best knows the mid-term and longer-term consequences of heart surgery, and we feel they are responsible for educating the rest of the stakeholders in the care of the patient, including psychosocial and mental health support as indicated. This strategy is precisely the model used for inpatient care, where one individual is not responsible for all aspects of optimal care (surgery, imaging, nutrition, nursing, anesthesia, etc.); rather, a team is assembled to provide the best possible outcome utilizing the expertise of multiple different individuals. This strategy of care is the model we propose to bring to the ambulatory setting. In addition to educating the rest of the care team, members of the medical home team should provide the patient and family education in the ambulatory setting, to assist in real-time understanding of multiple topics:

- the cCHD,
- the interventions performed,
- the current cardiovascular assessment,
- the potential long-term consequences of the procedures, and
- the importance of transition to self care.

Models implementing advanced practice nursing after discharge from cCHD surgery have been shown to reduce maternal worry as well as improve outcomes of infants, and it is our expectation that similar benefits would be seen over time in the ambulatory setting.[48]

Finally, it is our hope that this model

- will stimulate discussion and information sharing between centers, practitioners, patients, and families;
- be modified constantly over time, and
- potentially adopted by other invested parties such as the Society of Thoracic Surgeons and European Association for Cardio-Thoracic Surgery.

These groups have collaborated for a number of years to create very large databases to examine short-term outcomes. Their results have been impressive.[49-52] Adding a longer-term care module will allow improved feedback on surgical and in-hospital strategies of care and speed the process of collaborative learning across centers. Reliance solely on perioperative studies to improve outcomes is helpful but short sighted. Similarly, reliance on outpatient cardiology appointments with highly variable follow-up strategies to determine the "success" of cardiac surgery for cCHD is inconsistent and incomplete; ambulatory strategies must evolve for continued improvement in outcomes. Tens of thousands of data points are being wasted.

We anticipate that

- some will disagree with this strategy,
- additional technology and/or later findings will change these recommendations,
- many (if not most) will disagree with the exact type and timing of testing, and
- implementation will necessarily be variable across centers and geography depending upon interest and resources.

To our knowledge, routine, standardized follow-up protocols do not exist for most cCHD, although there is increasing interest in standardization for patients with a univentricular heart, including at institutions such as Children's Healthcare of Atlanta (Mahle W, personal communication), Lucille Packard Children's Hospital (Wright G, personal communication), and undoubtedly others. Much of what we have learned in this regard is based on the recognition of multi-system consequences of the Fontan procedure from the seminal work from the Single Ventricle Survivorship Program at The Children's Hospital of Philadelphia.[53-58] We speculate that other forms of cCHD, particularly with residual physiologic perturbations such as in tetralogy of Fallot, may also have as-yet-to-be-defined multi-system consequences as well; only systematic follow-up of large cohorts of patients will identify these potential morbidities. Our roadmap put forth here for patients with a univentricular heart, while similar, is somewhat different than that recently proposed by Rychik,[7] both in

terms of frequency of testing and by non-inclusion of standardized invasive testing. Which protocol is "right", most cost-effective, or likely to become standardized remains to be seen. However, we must start somewhere.

Finally, we wish to emphasize that comprehensive follow-up strategies should not be limited to those patients with univentricular hearts. We speculate that other forms of cCHD – particularly those with residual physiologic perturbations such as chronic semilunar or atrioventricular valve regurgitation – may also have yet-to-be-defined multi-system consequences. Only systematic follow-up of large cohorts of patients will identify these potential morbidities. The authors, as well as the editors of Cardiology in the Young, welcome contrary and conflicting opinions, and look forward to the dialogue.

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Figure Legends and Tables

Table 1 – Important longer-term consequences of selected cardiac surgical procedures in the neonate and infant

All

- Unplanned interventions
- Chronotropic impairment
- Decreased exercise performance
- Obesity
- Risk of bacterial endocarditis
- Restrictive lung disease
- Recurrent laryngeal nerve injury
- Neurodevelopmental delay
- Psychosocial maladjustment
- Genetic co-morbidities (if present)

Neonatal Surgery

Arterial Switch Operation

- Supravalvular pulmonary stenosis and branch pulmonary artery narrowing
- Neo-aortic valve regurgitation
- Neo-aortic root dilation
- Coronary obstruction or occlusion
- Pulmonary hypertension
- Residual septal defects

Repair of arch obstruction/interruption with ventricular septal defect

- Residual septal defects
- Left ventricular outflow tract obstruction
- Residual arch obstruction

Repair of common arterial trunk

- Neo-aortic valve stenosis or regurgitation
- Neo-aortic root dilation
- Conduit obstruction
- Branch pulmonary artery narrowing
- Pulmonary hypertension
- Residual septal defects

Repair of totally anomalous pulmonary venous return

- Pulmonary venous obstruction
- Pulmonary hypertension

Infant surgery

Repair of atrioventricular septal defect

- Residual atrial or ventricular septal defect
- Residual atrioventricular valve regurgitation or stenosis
- Left ventricular outflow tract obstruction

- Pulmonary hypertension

Repair of tetralogy of Fallot

- Right ventricular outflow tract obstruction
- Pulmonary regurgitation
- Branch pulmonary artery narrowing
- Aortic root dilation
- Aortic regurgitation
- Atrial and ventricular arrhythmia

Staged reconstruction for functionally univentricular heart (Fontan procedure)

- Pulmonary artery narrowing
- Ventricular dysfunction
- Atrioventricular valve regurgitation or stenosis
- Veno-venous collaterals
- Aortopulmonary collaterals
- Ventricular outflow obstruction
- Residual arch obstruction
- Pulmonary arteriovenous malformations
- Atrial arrhythmias
- Neo-aortic root dilation and regurgitation
- Conduit or venous pathway obstruction
- Hypercoagulability
- Plastic bronchitis
- Altered bone density
- Short stature and delayed puberty
- Cirrhosis
- Protein losing enteropathy
- Esophageal varicies
- Peripheral venous stasis and varicies

Table 2 – Survey respondents (n=434)

Location		
United States		
• Northeast	104	(24.4%)
• Mid-Atlantic	44	(10.1%)
• Southeast	85	(19.8%)
• Midwest	54	(12.4%)
• South	16	(3.7%)
• Northwest	16	(3.7%)
• West	41	(9.4%)
Outside United States	74	(17.1%)
Practice Type		
• Hospital Faculty	266	(58.6%)
• Private Practice	135	(31.1%)
• Fellowship	33	(7.3%)
Years in Practice		
• Fellow	31	(7.3%)
• <5 years	72	(15.9%)
• 5-10 years	78	(17.2%)
• 10-15 years	82	(18.1%)
• 15-20 years	70	(15.4%)
• >20 years	101	(23.3%)
Average Number of Outpatients per Week		
• <10	58	(13.4%)
• 10-20	180	(41.5%)
• 21-30	105	(24.2%)
• >30	91	(21.0%)

Figure Legends

1A-G. Results from an internet survey conducted in 2006 regarding frequency of diagnostic testing following surgery for tetralogy of Fallot, the arterial switch operation and Fontan palliation for functionally univentricular heart. See text for details.

2. Schematic representation of "The Roadmap".

Abbreviations in appendix:

CT=computerized tomography

ECG=electrocardiogram

MRI=Magnetic Resonance Imaging