Updates in Congenital Heart Disease: Can Outcomes Improve?

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Abstract

Keywords

► congenital heart disease
► quality improvement
► pulse oximetry screening

Despite numerous advances in medical and surgical management, congenital heart disease (CHD) remains the number one cause of death in the first year of life from congenital malformations. The current strategies used to approach improving outcomes in CHD are varied. This article will discuss the recent impact of pulse oximetry screening for critical CHD, describe the contributions of advanced cardiac imaging in the neonate with CHD, and highlight the growing importance of quality improvement and safety programs in the cardiac intensive care unit.

Improved Detection of Critical Congenital Heart Disease

The late detection of critical CHD has been recognized as a contributor to the early morbidity and mortality associated with CHD.4,5 Earlier critical CHD diagnosis in a New Zealand study showed 16% mortality compared with 27% mortality for infants with critical CHD who received a late diagnosis.6 Despite improvements in the prenatal detection of CHD, the rates of prenatal detection have shown significant variation between hospitals and disappointing national detection rates for transposition of the great arteries (37%) and coarctation of the aorta (34%).7 Because of the difficulty in detecting mild and even moderate cyanosis, physical examination has also shown only a 50% likelihood of detecting critical CHD.

National recommendations for pulse oximetry screening for critical CHD are becoming more common (►Table 1). A Cochrane analysis of pulse oximetry screening for critical CHD showed moderate sensitivity and high specificity.8 In the United States, newborn screening has been adopted in all 50 states and the District of Columbia.9 A recent study has shown the association of state implementation of pulse oximetry screening policies with a significant reduction in early infant cardiac deaths.10 States with legislation mandating newborn screening achieved a 33% reduction in early cardiac deaths due to CCHD compared with states with no policy or a nonmandatory policy. This reduction in mortality resulted in approximately 120 lives saved (95% confidence interval [CI]: 38–181) per year and an additional 21% reduction in other/unspecified CHD deaths in states, equivalent to another 117 lives saved (95% CI: 38–185) per year. Identifying infants with critical CHD early allows them to receive timely interventions, such as cardiovascular surgical repair or cardiac catheterization, and dramatically improves their chances of avoiding not only death but also morbidity associated with heart failure. Improvements in screening
Cardiac imaging in this patient population.

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Table 1 Global critical congenital heart disease screening implementation

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<th>National recommendation to screen</th>
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*National recommendation to screen includes countries where a government policy or a professional medical society recommends or requires screening for critical congenital heart disease.

during the neonatal phase, whether through pulse oximetry
screening, prenatal ultrasound and postnatal exam, can all
impact early identification of this disease.

**Advanced Cardiac Imaging in the Neonate with Congenital Heart Disease**

The development of two-dimensional echocardiography in
the 1960s revolutionized the approach to the newborn with
CHD and has become the mainstay for diagnosis and man-
agement of these patients. However, echocardiography does
have restrictions, such as the need to acquire images from
certain “anatomic windows,” difficulty demonstrating cer-
tain three-dimensional relationships, and limited ability
to provide hemodynamic information. Over the past three
decades, advancement in alternative imaging modalities,
particularly cardiac magnetic resonance imaging (CMR) and
computed tomography (CT), have continued to improve
and revolutionize the approach to the diagnostic and hemo-
dynamic evaluation of patients with CHD. Efforts have
also progressed through multiple societies and organiza-
tions to develop guidelines for the appropriate use of advanced
cardiac imaging in this patient population. As advance-
ments continue and indications become more well defined,
newborns and infants with CHD benefit from advanced
cardiac imaging techniques to improve diagnostic accuracy
and provide a broader spectrum of noninvasive hemody-
namic evaluation, while at the same time decreasing safety
concerns associated with these techniques.

Advantages of advanced cardiac imaging vary based on
the modality, but both CMR and CT are noninvasive and
provide images with large field of view that is unconstrained
by acquisition windows or imaging planes. Both techniques
have the ability to provide functional information (chamber
volume, stroke volume, ejection fraction) of the heart based
on acquired three-dimensional data that is not reliant on
modeling or anatomic assumptions. Likewise, both modal-
ities can provide information on myocardial perfusion and
tissue viability. CMR is particularly advantageous because it
is radiation free, provides excellent tissue contrast and
temporal resolution, allows for blood flow analysis (velocity
and volume quantification), as well as tissue characteriza-
tion. An additional benefit of CMR is that significant diag-
nostic information can be obtained without the use of con-
tact agents. CT, on the other hand, provides very high
spatial resolution and can be performed very quickly, albeit
with the need for intravascular contrast in most cases.
Cardiac CT does utilize radiation, but advancements in
scanner technology have drastically reduced dosages such
that modern cardiac CT scans can be obtained with similar or
less radiation than is required for a typical chest X-ray. Both
advanced cardiac imaging modalities also have traditionally
shared some common disadvantages, namely, being intoler-
ant to patient motion, requiring sedation or breath holds in
many cases, and being difficult to perform in patients with
cardiac arrhythmias. The techniques are also technically
complex, requiring specialized equipment as well as expert-
tise in the field. Nonetheless, advanced cardiac imaging has
significantly strengthened the noninvasive diagnostic capabilities for cardiovascular imaging over the past two decades. Developments such as motion-corrected imaging sequences, rapid imaging scanners and sequences, neonatal-specific MRI coils, and the use of novel clinical approaches such as “feed and sleep” imaging have all contributed to the increased applicability of CMR and cardiac CT in management of the neonate with CHD.

Although echocardiography is able to provide excellent diagnostic details in most newborns with CHD, there are questions that can sometimes remain after a study is performed. In some instances, questions regarding the three-dimensional relationship of heart structures can remain, such as the ventriculoarterial connection relationship in complex heart disease or anatomic details of atrioventricular valves in the setting of questionable ventricular hypoplasia. These details can impact the approach to intervention and are important to obtain before repair is undertaken (► Fig. 1).

Echocardiography can also sometimes leave questions regarding anatomic details of vascular anatomy or its relationship to other structures within the chest. Advanced cardiac imaging plays an important role in the evaluation of vascular rings (► Fig. 2), determining the presence or absence of aortopulmonary collaterals in certain diseases, visualizing details of venous anatomy related to heterotaxy syndrome, and defining structural details of anomalous pulmonary veins (► Fig. 3). Additionally, echocardiography is limited in its ability to provide some details that impact hemodynamics and physiology. For example, volume quantification in the setting of ventricular hypoplasia to determine adequacy for a biventricular repair, or measurement of pulmonary-to-systemic blood flow ratio or differential pulmonary blood flow in certain disease states can guide the management approach (► Fig. 4). CMR, in particular, can be an invaluable tool for defining the nature of intracardiac or thoracic masses due to its ability to provide tissue...
characterization information which can guide medical decision making (►Fig. 5).

Advanced cardiac imaging currently plays an important role in answering these questions in a noninvasive and reliable manner, and continued technical advancements will only strengthen the ability to provide detailed anatomic and functional imaging while minimizing or eliminating safety concerns associated with sedation, contrast, and radiation exposure.

Quality Improvement and Safety Programs in the Cardiac Intensive Care Unit

Patient care in the CICU is increasingly complex and requires a well-coordinated team with several disciplines including intensive care physicians, anesthesiologists, nursing, cardiologists, dieticians, social workers, perfusionists, respiratory technicians, fellows, and surgeons. Care coordination between these providers is essential to optimize care delivery and outcomes.

With growing emphasis on public reporting, data transparency, and patient advocacy, understanding ways to improve care coordination and outcomes is essential. Quality improvement science is the systematic study of processes and systems involved in care delivery to optimize outcomes. Principles of quality improvement can be applied to a single center and are also well suited to multi-institutional collaboratives. To optimize the success of this work, multidisciplinary collaboration is also essential.

Randomized controlled trials and large multicenter drug trials are becoming less common and more difficult to conduct due to cost, regulatory oversight and inherent heterogeneity of CHD. Significant variation in outcomes remains across hospitals, and there is a need to benchmark quality and reduce morbidity and mortality. Because of the limitations of large multicenter trials, quality improvement collaboratives and large database registry studies are important platforms. Large databases such as the Society of Thoracic Surgeons, Pediatric Cardiac Critical Care Consortium (PC4), and National Pediatric Cardiology Quality Improvement Collaborative (NPCQIC) are examples of population level data registries which allow for the compilation of large amounts of patient demographic, care, and outcomes data. The PC4\(^{19}\) began in 2009 and sought to improve outcomes for patients with critical pediatric and CHD using continuous quality improvement through data analysis and collaborative learning.

Further, PC4 and NPCQIC are focused on quality improvement and outcomes, and recent studies have demonstrated the potential impact of conducting research and quality improvement utilizing these large multicenter registries.\(^{20,21}\) The NPCQIC is a multicenter collaborative focused on reducing mortality and improving weight gain between the first- and second-stage palliative procedures in patients with hypoplastic left heart syndrome. Anderson et al demonstrated a 44% reduction in mortality and a 28% reduction in interstage growth failure through the NPCQIC with particular focus on “all teach, all learn” information and data sharing as well as parental engagement.\(^{20}\) The success of registries such as PC4 and NPCQIC clearly highlights distinct benefits of timely feedback on performance and outcomes, maintaining and tracking practice variation and linking these to outcomes, and importantly to facilitate wide-scale learning on rare diagnosis, such as hypoplastic left heart syndrome.

Fig. 2  Fetal echocardiogram demonstrated probable right aortic arch with a left-sided ductus arteriosus (A). Postnatal cardiac MRI performed at 6 weeks of age verified the presence of a right aortic arch with an aberrant left subclavian artery and Kommerell’s diverticulum, confirming the presence of a vascular ring (B). The confirmatory imaging was accomplished without sedation using a feed and bundle technique and did not expose the newborn to ionizing radiation. MRI, magnetic resonance imaging.
Multicenter data aggregation allows participating centers to amass enough patients to study demographics, practice patterns, and analyze outcomes in a manner which would be impossible using only single-center data. Ultimately, large clinical registries should inform quality improvement to drive improvement in outcomes. Using this model for quality improvement, PC4 has recently launched its first multicenter quality improvement initiative to reduce cardiac arrests in patients in the CICU. The project is a multi-institutional study based on aggregate PC4 data using an evidence-based and empirically derived care bundle. Although data are still being collected and analyzed, early analyses indicate important reductions in cardiac arrest rates. Studies and quality improvement efforts such as these will improve outcomes, spread change, and improve our collective understanding of this complex patient population.

While large registries provide exceptional opportunities for understanding demographics and outcomes, single-center quality improvement remains an important strategy to improve outcomes locally. Successes from implementing this approach have been seen in improving and sustaining hand-off processes between the cardiovascular operating room and CICU, where performance improvement methodology and key drivers developed with input from many disciplines allowed clinical staff to improve communication, technology, and information transfer and sustain a shared mental model related to the patients’ clinical condition and plan of care.

Translating scientific discovery and advances in clinical care to optimize care for both long- and short-term outcomes requires a coordinated effort by multiple stakeholders. A recent expansion of databases and registries has created a necessity to streamline data collection, analytics, and outcomes reporting to simplify research, quality improvement, and patient advocacy. Cardiac Networks United has been launched to integrate pediatric and congenital cardiovascular research and quality improvement networks and databases. This will allow researchers to link practices and outcomes across the lifespan for congenital heart patients as well as allow clinical providers to integrate new findings.
and lessons learned from both research as well as focused quality improvement networks. Through the evolution of efforts such as Cardiac Networks United, multicenter quality improvement and data sharing will continue to advance the care of children with critical CHD.

Conflict of Interest
None declared.

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