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Preface

Our nation's health care infrastructure faces unprecedented challenges in the face of the COVID-19 pandemic, and the congenital heart disease (CHD) community is no exception. These challenges include looming resource scarcities of equipment, personnel, and blood. In addition, there are the substantial infection risks to patients, family members and staff. These factors necessitate thoughtful but often difficult decisions on how to best triage patients with CHD. Our relatively small workforce adds another dimension to the challenge, since the rapid spread of COVID-19 could result in program closure at a moment's notice secondary to insufficient personnel from infection or quarantine. While many sectors of our society can be placed on hiatus during this period of crisis, our patients' diseases continue requiring care, particularly amongst newborns and infants who often require surgery during a narrow temporal window for satisfactory outcomes. Practitioners are tasked with optimizing care in the presence of the current and rapidly changing circumstances. While statements have been published relative to adults undergoing surgery,¹ guidance with respect to congenital heart disease patients is currently lacking and is the goal of this manuscript. As we have seen in other countries facing this pandemic, the thoughtful allocation of resources is paramount to the overall welfare of the community ^{2,3} and has led to such strategies in our country.^{4,5} The safety of our patients, healthcare providers, and our communities is our chief concern.

This document is not meant to be a guideline but is designed to provide *guidance* for decision-making as we face unparalleled challenges related to congenital cardiac surgery care during this pandemic. The circumstances are rapidly changing, even hourly, therefore, the principles outlined herein are meant to be fluid and adaptable. They should be continually reappraised in the context of the dynamic circumstances within a given institution, population base

and geographic location. These principles can also provide a framework for prioritization of surgery in other situations when there is lack of resources or personnel, or both. This document is not meant to be prescriptive, but rather serves as an outline of guiding principles to be interpreted in a particular context. Lastly, while individual anomalies may be specified, each patient should be considered individually in the context of their clinical status, disease state, institution, and community.

Perhaps more than ever, these times require us as a specialty, albeit small but with a valuable scope of skillsets, to *collaborate and cooperate*. This includes the sharing of knowledge and resources, patient transfer in selected situations, and communicating frequently amongst ourselves to provide emotional support and mental fortitude during these periods of stress and isolation.

Congenital heart surgeons have a track record of effectively working together to advance the specialty, promote quality training and education of our surgical community, and through bold innovation solve some of the most difficult clinical challenges. We are now confronted with a different kind of challenge – a public healthcare crisis. We can rise to meet this challenge by utilizing our collaborative and intellectual abilities to problem solve, plan and prevail for the benefit of children and adults suffering from CHD. We must work together with strong leadership from our institutions and central, shared decision-making teams to handle the influx of issues and concerns related to the pandemic.

Triage of Surgical Patients

In the setting of substantially limited resources, proper prioritization of patients requiring surgery is of paramount importance. To date, the biggest difficulties have not been COVID

infections in CHD patients, which have been rare. The biggest threat to our patients is the sudden lack of resources, including the requisition of ORs (transformed into intensive care units), ventilators and healthcare providers for the fight against COVID. As a result, the surgical schedule must be culled to only the most urgent cases, which in our specialty is very difficult with many gray zones. Many factors pertaining to an individual case must be weighed, including: 1) resource-utilization, such as anticipated ventilator duration, intensive care unit stay, blood product usage, and other supplies that are or may become limited, 2) clinical status of the patient and risk of delaying surgery, 3) risk of exposure for the patient, family, and healthcare staff, and 4) co-morbidities and complexity of the procedure with implications on the usage of hospital resources, 5) in teaching hospitals, training may have to be curtailed and the most experienced surgeons used liberally, and 6) the safety of the patient's social and clinical situation if surgery is delayed. These decisions will often be made in the context of a hospital that is weighing the needs of other patients from different specialties with similar needs.

The timing of surgery is determined by a variety of factors – the clinical status of the patient, recommendations of the CDC, government officials, and professional societies (e.g., American College of Surgeons), and local hospital protocols. Importantly, this is a shared decision-making with the patient, family, and other specialists with transparency regarding the risks and benefits of proceeding with surgery versus waiting. The difficulty of this decision-making regarding timing is compounded by the realization that the duration of this pandemic and thus the delay for a given patient remains unknown.

Table 1 is provided as *general guidance* regarding the timing of intervention for various diagnoses, depending on clinical status and other factors. It is not exhaustive and not meant to be a guideline, but highlights common conditions and scenarios. As stated earlier, this should be

interpreted with flexibility depending on the current state of resources within a given institution and community. It is important to note that ongoing, close cooperation and communication between the surgical and medical teams is essential to ensure an accurate diagnosis and effective treatment strategy with the fewest resources and involved personnel. While in the setting of unrestricted resources certain conditions can be managed in a standard fashion, consideration during this time should be given to alternative approaches that may drain less resources and/or result in shorter hospitalization; e.g., a ductal stent may optimize resource utilization for a given institution as opposed to a surgical systemic-to-pulmonary artery shunt. Depending on the impact of this pandemic, there may be circumstances in the near future when it may not be appropriate or feasible to offer interventions to high-risk patients requiring high resource utilization with anticipated poor outcomes.

Preserving the Workforce and Regional Collaboration

Given the small size of each institutional workforce practicing congenital cardiac surgery (median of 3 surgeons per practice in the United States),⁶ which possess unique skillsets not replaceable by other providers, strategies to maintain the integrity of the workforce are crucial. Institutions may re-deploy members of the congenital cardiac surgery team to other patient-care settings, further depleting resources and also increasing exposure. In response, employing workplace strategies to reduce overall exposure, such as one-week on/one-week off rotation schedule for selected health-care providers, appropriate use of personal protection equipment (PPE), and downsizing of clinical teams and worksites, seems prudent. Vigilant surveillance for symptoms among team members is critical given the close proximity of congenital cardiac team members working together; one exposed or positive provider can put the entire team and

program at risk because of quarantine requirements, resulting in a program being shut down because of insufficient staff to provide care. Precautions should also be taken for non-clinical support staff to minimize their exposure. Strategies of remote tele-working and rotational schedules should be applied to all staff when feasible.

Regional *collaboration* should be considered, with the realization that small programs are at risk for closing in the setting of infected staff members and/or those requiring quarantine. While neighboring congenital cardiac surgery programs may have had a history of competition, the importance and necessity of cooperation and collaboration cannot be overemphasized during this time of public healthcare crisis. *A patient-centric mindset should be the overarching emphasis* and programs and practitioners should find ways to work together and support each other. Discussions could include strategies for reciprocal hospital privileging that allow staff to move from one site to the other if needed, sharing COVID-related perioperative protocols or other newly created institutional care strategies and crisis management guidelines that respond to the changing needs of COVID-19. Additionally, critically ill patients at institutions facing severely depleted resources may be best served by transferring to institutions with more plentiful resources.

Surgical Consultation and Clinic Visits

Babies will continue to be born each day and the 1% incidence of congenital heart disease will remain unchanged during this time of crisis. Evaluation and early surgery for many children must continue while much of the world is on hold. An adaptive clinical model should be applied to minimize exposure and prioritize patients for surgery during this crisis period. Differentiating who requires surgical evaluation for potential intervention during this time and who does not can

be accomplished with a careful history, virtual visits, and review of diagnostic laboratory tests and imaging. After a multi-disciplinary discussion, further imaging or lab-work can be obtained as needed to allow for accurate risk stratification and surgical planning.

COVID-19 Exposure and Social Distancing

An ongoing concern is the relative risk of hospital exposure to COVID-19. While each hospital strives to carefully monitor and appropriately care for COVID-19 patients, in-house patients and healthcare providers may be positive prior to symptom onset or knowledge of their status, putting incoming surgical patients and their families at risk. Furthermore, many of our patients' family members may be residing in group facilities, e.g., Ronald McDonald House, also placing them at risk for exposure. While the magnitude of such potential risk is unknown, it must be weighed against the urgency of the operation. COVID-19 has had the biggest clinical impact on older patients, especially those with predisposing conditions; however, younger patients without medical conditions are being affected and have also died. This finding is an important consideration since the congenital cardiac population often has underlying pathophysiological abnormalities and co-morbidities that place them at higher risk in the setting of a respiratory illness.

Social distancing can be particularly challenging for our patients, given their age and family structure. Many of our patients have multiple siblings, each with other potential exposures, and depending on their age, require supervision or care by adults. Adequate psychosocial support for the child and their family must also be considered, while being mindful of the risk of COVID-19 exposure and spread. Individual decision-making must be considered in this setting, taking into account specific hospital policies and community recommendations so

exposure is minimized; e.g., one parent bedside during hospital stay as opposed to additional family members. The adverse psychosocial impact of such a strategy cannot be underestimated and alternative support mechanisms be provided.

Discussions continue regarding the appropriate use of personal protection equipment (PPE) for healthcare workers and screening of incoming patients to prevent spread of COVID-19 within the hospital setting. Such policies will inevitably continue to change with PPE supply, evolution of testing, and knowledge regarding the spread of the virus.

Ideally, during this pandemic preoperative COVID-19 testing should be performed on every patient, even asymptomatic patients, as well as their parents. While the testing yield is poor in asymptomatic patients early in the disease before viral shedding,⁷ a positive test would substantially change both decision-making regarding their surgery as well as precautions taken during their hospitalization, use of limited PPE, and interaction with providers and family members. How to manage a COVID-19 positive patient with CHD who requires urgent surgery remains unclear since the effects of cardiopulmonary bypass and mechanical ventilation on an asymptomatic or symptomatic COVID-19 positive patient are currently unknown.

Role of Congenital Cardiac Surgeons in the Care of COVID-19 Positive Non-Cardiac Patients

While our priority and focus has centered on patients with congenital heart disease, congenital cardiac surgeons also play an integral role in the care of COVID-19 positive patients without cardiac disease. This primarily includes patients with respiratory failure from COVID-19 who may require extracorporeal membrane oxygenation (ECMO) support. Although infrequently needed for COVID-19 positive children thus far, the age range of COVID-affected

patients continues to evolve with an increasing number of younger patients being affected. Institution of pediatric ECMO is a unique skill that is most often implemented by congenital cardiac surgeons. While participation of the surgeon is often mostly technical, i.e., cannulation, it also includes patient selection guidelines, strategies for ECMO application with the fewest personnel, transportation protocols, post-ECMO limb monitoring, chest washouts, etc. Early consultation with the surgical service and coordinated efforts with clear communication between the involved personnel – surgery, critical care, perfusion, nursing, etc., cannot be overemphasized since the potential for controversy and disagreement is more likely in the COVID-19 setting. Strategies to limit healthcare personnel exposure during the institution of ECMO and the care of COVID-19 patients on ECMO need also be employed.

The decision on whether to employ ECMO for COVID-19 positive patients can be controversial and will depend on resource availability, co-morbidities, and pre-illness clinical status. ECMO should be considered in a pediatric patient who is positive and otherwise healthy prior to COVID infection in an institution with adequate resources and infrastructure.

Training and Education

Surgical trainees in the field of congenital heart disease are also severely impacted by the current pandemic. Residents and fellows are frequently the frontline for many urgent in-patient emergencies, e.g., cardiac arrest in post-cardiac surgery patients, ECMO cannulation, placement of central venous or arterial monitoring lines, intubation, etc. They are also on a specific and very short timeline for acquiring surgical experience with case number and procedural requirements. However, this must be balanced with the risk of their exposure and well-being. Hospital policies may limit their participation in in-patient activities, and some may require

periods of quarantine. The cumulative effect of these issues could result in a markedly reduced training experience, thus compromising their ability to fulfill certification metrics. Most congenital cardiac surgery residents have already completed 5-9 years of elite thoracic surgical training in order to spend 1-2 precious years of training at one of the few congenital residency programs in the country. While post-graduate medical and surgical education has been complemented by various remote or on-line learning modules over the last decade, the bulk of *surgical* learning continues to be centered in the operating room and ICU setting – this experience is essential and not replaceable with on-line education. While some flexibility during this crisis will be considered by the ACGME and ABTS, there may be circumstances where extension of the duration of training is needed; governing Boards and Program Director input will be necessary to ensure competence of the graduating trainee. In the meantime, efforts need to be made to minimize unnecessary exposure to infection, take appropriate precautions, and maximize all supplemental educational measures, e.g., video conferencing, virtual classrooms, etc., so trainees don't fall too far behind.

Mental Toughness and Emotional Stability

The history of our specialty is defined by courage, resiliency, and tenacity. Congenital heart surgeons complete one of the longest and most rigorous training regimens in medicine. It is a specialty marked by long and technically difficult operations. It is a specialty of high risk and high reward, and the cardiac surgical personality thrives on it. Difficult decision-making and critical conversations with colleagues and family members are part of our daily experience. As leaders of our teams, we manage pressure and work to address and minimize our team's fatigue,

anxiety, or moral distress. These attributes have never been at a higher premium than at the present moment.

During this pandemic, the difficult decision-making has shifted to an unprecedented realm – who gets surgery and when, how limited resources will be used, and how our team will be allocated toward the overall good. With our depth of experience and reservoir of emotional and mental toughness, we as a community are a critical public health resource. Our patients, families, institutions, and regions are depending upon us to help them navigate these difficult circumstances. This historic challenge calls on us to, as we have throughout our history, meet the toughest of problems head-on.

We are a small community with a rich network of relationships. We are now trying to find ways to *work together and uplift one another while under duress*. While isolated physically, we are virtually connected; and this support is vital to our success. Frequent and intentional phone calls, sharing personal experiences, and reaching out to colleagues if institutional capabilities are threatened, as well as finding other ways to balance personal and professional life, are necessary to overcome the present challenge.

Conclusion

Unprecedented times call for unprecedented measures. Prioritization and appropriate timing of surgery are necessary at this time. Practical guidance strategies range from ensuring safety and tactics for specific lesions of the patients, to maintaining emotional stability of the staff. Our specialty has been marked by solidarity and camaraderie, and carries a history notable for

collaboration, flexibility, adaptation and instant readiness. The time to execute these qualities is here and now.

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Table 1. Congenital Lesions and Surgical Prioritization.

| | | Emergent (24-48 hours of diagnosis when adequate resources) | Urgent (within 1-2 weeks when adequate resources) | High priority elective (>2 weeks when adequate resources) |
|---------------|-----------------------------|--|---|--|
| <u>eonate</u> | | note: timing for categories will deper pending cases | nd on resources available, ins | titutional protocols, and other |
| Shunts, | Mixing Lesions | | | |
| | TAPVC/cor | obstructed | increasing gradient | |
| | triatriatum TGA | | <1 week if IVS | 2-4 weeks if VSD |
| | Truncus Arteriosus | | | if stable |
| | Tetralogy of | severe hypoxemia/hypercyanotic | symptomatic | |
| _ | Fallot | spells | | |
| Regurgi | tant Lesions | | | |
| | Ebstein Anomaly | | refractory medical manage | ment |
| Obstruc | tive Lesions | | | |
| | Coarctation | shock unable to stabilize on PGE | if able to stabilize on PGE | |
| | Critical Aortic Stenosis | shock unable to stabilize on PGE | if able to stabilize on PGE | |
| PGE-de | pendent pulmonary b | blood flow | | |
| | PA/IVS | | if PDA stent not available | |
| PGE-dep | pendent systemic blo | od flow | · | |
| | HLHS | intact, restrictive atrial sepum if BAS not available | case and surgeon dependent | case and surgeon dependent |
| Other | | | | |
| | Shunt | shunt thrombosis | shunt stenosis | |
| | Arrhythmias | symptomatic congenital heart block | unable to medically manage/ | externally pace |
| | ALCAPA | once medically stabilized | | |
| fant | | | | |
| Shunts I | eft> Right | | | |
| | VSD | | symptomatic CHF on medical mgmt | failure to thrive |
| Shunts I | Right> Left | | · | · |
| | Tetralogy of Fallot | | symptomatic (spells, cyano | sis) on medical mgmt |
| Regurgi | tant lesions | | <u> </u> | |
| | AVSD | | | Tri21 with pulmonary overcirculation, consider age or patient to optimize repair, significant regurgitation unable to manage medically |
| | Ebstein Anomaly | | | increasing right sided heart failure on medical mgmt |

| | Mitral Regurgitation | | | symptomatic CHF on medical mgmt |
|---------|----------------------|---------------------------------|---|------------------------------------|
| | Aortic | acute, hemodynamically unstable | I | enlarging LV, decreasing LV EF, |
| | Regurgitation | | | symptoms |
| Obstruc | tive Lesions | | | |
| | Valve prosthesis | thrombosed prosthesis | | |
| | AS/LVOTO | | | decreasing LV EF, symptoms |
| | RVOTO | | | decreased RV function |

Other

| Shunt | shunt thrombosis | shunt stenosis | |
|-----------------|------------------|--------------------------|--|
| DCM/HF | | CHF failing medical mgmt | failure to thrive |
| BDCPA candidate | | 6 | increasing cyanosis with current shunt, shunt stenosis |

<u>Children</u>

| Regurgit | tant lesions | | | |
|----------|------------------------------|---|---|--|
| | Mitral Regurgitatio | n | <u>`</u> Q` | symptomatic CHF on medical mgmt |
| | Aortic Regurgitation | acute, hemodynamically unstable | | enlarging LV, decreasing fxn, symptoms |
| Obstruc | tive Lesions | | | |
| | AS/LVOTO | | v | decreasing LV fxn, symptoms |
| | Valve prosthesis | thrombosed prosthesis | | |
| | RV-PA conduit obstruction | severe stenosis with severe RV dysfunction and/or ventricular arrhythmias | severe stenosis with RV dysfunction and/or systemic RV pressure | worsening right-sided failure |

Other

| DCM/HF | | CHF failing medical mgmt | failure to thrive |
|--|---|--|--------------------------------|
| Fontan candidate | | | increasing cyanosis |
| Endocarditis | cardiogenic or septic shock despite max medical mgmt | hemodynamcially stable, but uncontrolled infection | per guidelines |
| AAOCA | recent cardiac arrest, hemodynamically unstable, on mechanica support | history of aborted sudden of exertion | leath, chest pain with minimal |
| Combined lesions (i.e. MR and subAS) | hemodynamic compromise | moderate/severe individua | l lesions |

Adult Congenital

Regurgitant Lesions

| | | 1 | |
|----------|----------------------|---------------------------------|---|
| | Ebstein/TR | | increasing right sided heart failure on medical mgmt |
| | Mitral Regurgitation | | symptomatic CHF on medical mgmt |
| | Aortic | acute, hemodynamically unstable | enlarging LV, decreasing LV EF, |
| | Regurgitation | | symptoms |
| Obstruct | tive Lesions | | |

| | НСМ | | syncope/presyncope | |
|-------|------------------------------|---|---|-------------------------------|
| | Aortic Stenosis | | | decreasing LV EF, symptoms |
| | RV-PA conduit obstruction | severe stenosis with severe RV dysfunction and/or ventricular arrhythmias | severe stenosis with RV dysfunction and/or systemic RV pressure | worsening right-sided failure |
| Other | | | | |
| | Endocarditis | cardiogenic or septic shock despite max medical mgmt | hemodynamcially stable, but uncontrolled infection | per guidelines |
| | AAOCA | recent cardiac arrest, hemodynamically unstable, on mechanical support | history of aborted sudden death | |

Lesions are listed under the age-group(s) in which they most commonly present. Not included in this table is orthotopic heart transplantation. The decision to accept a donor heart during this pandemic depends on the clinical status of the recipient, the estimated risk of the donor's potential exposure to COVID-19 in their community and hospital, and the prevalence of COVID-19 in the hospital and community of the recipient in light of the immunosuppression the recipient will receive.

TAPVC=total anomalous pulmonary venous connection; TGA=transposition of the great arteries; IVS=intact ventricular septum; VSD=ventricular septal defect; PGE=prostaglandin-E1; PA/IVS=pulmonary atresia/intact ventricular septum; PDA=patent ductus arteriosus; HLHS=hypoplastic left heart syndrome; ALCAPA=anomalous left coronary artery from pulmonary artery; CHF=congestive heart failure; mgmt=management; AVSD=atrioventricular septal defect; Tri21=trisomy 21; LV=left ventricle; EF=ejection fraction; RV=right ventricle; AS/LVOTO=aortic stenosis/left ventricular outflow tract obstruction; RVOTO=right ventricular outflow tract obstruction; DCM/HF=dilated cardiomyopathy/heart failure; BDCPA=bi-directional cavopulmonary anastomosis; AAOCA=anomalous aortic origin of the coronary arteries; TR=tricuspid regurgitation; HCM=hypertrophic cardiomyopathy

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