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REVIEW PAPER

From Other Journals: A Review of Recent Articles by Our Editorial Team

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Abstract
In this review, we provide a brief description of recently published articles addressing topics relevant to pediatric cardiologists. Our hope is to provide a summary of the latest articles published recently in other journals in our field. The articles address the extracardiac anomalies in fetuses with congenital heart disease, post COVID-19 vaccination myocarditis, the use of cardiac magnetic resonance after the Fontan operation, congenitally corrected transposition in adults, robitically assisted congenital heart surgery and the increased risk of congenital heart surgery in patients receiving tracheal surgery during the same admission.

Keywords COVID-19 vaccination · Myocarditis · Fontan · Congenitally corrected transposition · Tracheal surgery · Robotic congenital heart surgery

Abnormal Extracardiac Development in Fetuses with Congenital Heart Disease

Understanding the extent of extracardiac anomalies (ECA) in fetuses with congenital heart disease (CHD) can inform prenatal and postnatal decision making. Also this knowledge may improve our understanding of the developmental origins of different outcomes in patients with CHD. ECA including structural brain anomalies (SBA) can be evaluated using prenatal fetal magnetic resonance imaging (MRI). This study evaluated a large number of fetal MRI across gestation in fetuses with CHD to evaluate the rate and nature of ECA [1].

The study evaluated 429 fetuses with CHD who had a fetal MRI between 17–38 weeks of gestation. ECA and SBA were evaluated in each type of CHD and classified by gestational age < 25 and above 25 weeks. Fetuses with fetal arrhythmia without CHD, atrial septal defect (ASD), isolated left superior vena cava, myocardial hypertrophy, or with isolated aberrant right subclavian artery were excluded.

There were 243/429 (57%) with ECA and 109 (25%) with SBA. The most common ECA after SBA were extrafetal (21%) (including placental, umbilical cord, and amniotic fluid anomalies) followed by urogenital (11%). The patients with normal prenatal genetic testing had relatively similar rates of ECA (54%) and SBA (19%). The most common SBA were hindbrain-midbrain (11%), dorsal prosencephalon (10%) and abnormal cerebrospinal fluids (11%). The prevalence and pattern were not statistically different between early (< 25 weeks) and late MRI (46% vs 54%). Among fetuses without genetic anomalies, the highest ECA rate was in ventricular/atrial septal defects (84%), and the lowest in transposition of great arteries (21%). Of note there was a high prevalence of extrafetal/placental anomalies highlighting the role of the placenta in patients with CHD. The study concluded that fetal MRI shows a high prevalence of ECA and SBA in fetuses with CHD. Fetal MRI is superior to ultrasound in detecting brain anomalies. As the prognosis of CHD is dependent in many cases on ECA, fetal MRI may aid the accurate prenatal diagnosis of ECA in CHD and thus help prenatal counseling of families with CHD.

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Clinically Suspected Myocarditis Temporally Related to COVID-19 Vaccination in Adolescents and Young Adults

Understanding the prevalence and the clinical course of post COVID-19 vaccination presumed myocarditis is crucial to counsel patients regarding vaccination. This multi-institutional study collected data on patients below the age of 21 years with suspected myocarditis 30 days after COVID-19 vaccination. Lake Louise criteria were used for cardiac MRI. Myocarditis cases were classified as confirmed or probable based on the criteria of the center for disease control [2].

The study included 139 adolescents and young adults with 140 episodes of suspected myocarditis (39 confirmed and 91 probable) at the study 26 centers. The median age was 15.8 years (14.5–17 years). Most patients were male 91%, white 66% and Hispanic patients 21%. The majority of cases (94%) occurred after the Pfizer-BioNTech vaccine and 91% occurred after the second dose. The most common presentation was with chest pain in 99%. 26 patients required intensive care treatment and two patients needed inotropic support. No patient required ECMO support and no death was reported. The median hospital length of stay was 2 days and the longest was 10 days. All patients had elevated troponin, 70% had abnormal EKG and/or arrhythmia including 7 patients with non-sustained ventricular tachycardia, and 19% had decrease ejection fraction by echocardiogram. Among the 97 patients who had a cardiac MRI at a median of 5 days post symptoms, 76% had late gadolinium enhancement, 54% had myocardial edema while 51% met the Lake Louise criteria for myocarditis. All the patients who had decreased ejection fraction by echo had a follow up echocardiogram that showed normal function.

The study concluded that almost all patients with post vaccine myocarditis had relatively mild course with normalization of ventricular function on follow up. Future studies for long-term outcomes are needed.

Integrated Clinical and Magnetic Resonance Imaging Assessments Late After Fontan Operation

Cardiac MRI (CMR) is an important tool for evaluation and management for patients with single ventricular congenital heart disease late after the Fontan operation. This large study evaluated the integration of clinical and CMR variables to identify patients with worse outcomes after the Fontan operation [3].

This was a retrospective study from the Boston Children's Hospital database. The primary outcome was time to death or listing for heart transplantation. Cox regression was used for this analysis. The study included 416 patients after the Fontan operation (62% male) with a median age of 16 years (interquartile range of 11–23 years). Over a median follow up of 5.4 years, 57 patients (14%) experienced the composite outcome (46 death, 7 heart transplant and 4 heart transplant listing). Elevated ventricular end diastolic volume (EDVi) was the strongest risk factor for the composite outcome. A classification and regression tree (CART) analysis was done and showed that EDVi above 156 ml/BSA$^{1.3}$ is the threshold for ventricular dilation associated with worse outcomes. Among patients with dilated ventricles (EDVi above 156 ml/BSA$^{1.3}$), the second most important factor was decreased global circumferential strain. Among patients without ventricular dilation, New York Heart Association classification of III or IV associated with worse mortality followed by worse single ventricle global function index which is a measure of ventricular size, hypertrophy, and function.

The authors concluded that in this cohort late after the Fontan operation, ventricular dilation by CMR was the strongest predictor of worse outcomes along with other risk factors including worse circumferential strain, worse functional status, and worse global function index. CMR is an important tool for risk stratification in this patient population.

The Fate of Congenitally Corrected Transposition of the Great Arteries (ccTGA) Unoperated Before Adulthood

Some patients with ccTGA remain asymptomatic and remain undiagnosed or misdiagnosed, not needing a surgical intervention before adulthood. The systemic right ventricle (SV) and valve are exposed to high pressure load and therefore develop dysfunction or regurgitation with increasing age. Often, there are associated lesions with ccTGA affecting the outcomes. Authors in this study examined the outcomes, therapeutic strategies, and risk factors associated with ccTGA combined with cardiac lesions that remained not intervened until adulthood [4].

The authors reviewed 117 ccTGA patients over a span of 9 years from a single institutional database. Patients were classified into 3 groups: intact ventricular septum (IVS) had 44 patients, ventricular septal defect (VSD) group had 19 patients, and pulmonary stenosis (PS) group (pressure gradient > 35 mmHg) had 54 patients. Mean age of the patients was 36.3 + / − 13 years. The PS group had significantly less systemic AVV regurgitation and ventricular dysfunction compared to the other two groups and had

the highest systemic ventricle ejection fraction. From the first visit to last follow-up, systemic ventricular ejection fraction of unoperated ccTGA decreased significantly. 9 patients underwent surgery with no hospital mortality.

In the intact ventricular septum group, patients needing surgery had significantly more severe SAVV regurgitation (61.1% vs 30.8%, $P=0.05$), SV dysfunction (33.3% vs 0.0%, $P<0.001$), and higher SV end systolic dimensions (SVESD) (35.3 + / − 6.1 vs 29.8 + / − 5.0, $P=0.01$) than unoperated patients. Patients receiving systemic atrioventricular valve (SAVV) replacement/valvuloplasty had a significantly increased SV ejection fraction (SVEF) 3.4 + / − 9.4 to 59.7 + / − 5.5 ($P=0.03$) and statistically more freedom from death and transplant than unoperated (p = 0.049). In the ventricular septum defect group, there were no significant differences in severe SAVV regurgitation (62.5% vs 45.5%, $P=0.65$), SVEF (52.1 + / − 13.5 vs 59.6 + / − 11.9, $P=0.21$), SV dysfunction (25% vs 0.0%, $P=0.16$), or SVESD (30.5 + / − 4.1 vs 31.8 + / − 7.1, $P=0.69$) between operated and unoperated patients. At the last follow-up visit, operated patients had significantly less late severe SAVV regurgitation (12.5% vs 70.0%, $P=0.03$) and SVESD (28.5 + / − 2.3 vs 34.2 + / − 5.5, $P=0.03$) than unoperated patients. There was no significant difference in late SVEF (53.1 + / − 10.3 vs 50.1 + / − 8.3, $P<0.001$) between operated and unoperated patients.

In the PS group patients there were no significant differences in severe SAVV regurgitation (17.4% vs 6.5%, P = 0.38), SVEF (60.2 + / − 5.1 vs 63.0 + / − 5.5, $P=0.06$), SV dysfunction (0.0% vs 0.0%, $P>0.99$), or SVESD (20.0 + / − 5.8 vs 21.1 + / − 5.1, $P=0.51$) between operated and unoperated patients. At the last follow-up visit, there were no significant differences in late severe SAVV regurgitation (9.5% vs 6.5%, $P>0.99$), SVEF (59.4 + / − 8.0 vs 60.6 + / − 7.3, $P=0.58$), SV dysfunction (9.5% vs 6.5%, $P>0.99$) or SVESD (22.9 + / − 7.9 vs 20.8 + / − 6.0, $P=0.42$) between operated and unoperated patients. Multivariate analysis showed that severe SAVV regurgitation (hazard ratio [HR] 19.41, 95% confidence interval [CI] 5.04–74.72, $P<0.001$), SV dysfunction (HR 4.97, 95% CI 1.60–15.48, $P=0.01$), and physiologic repair (HR 5.12, 95% CI 1.37–19.18, $P=0.02$) were risk factors for mortality, transplant, and congestive heart failure.

The authors concluded that PS protects against systemic atrioventricular valve regurgitation and ventricular dysfunction. Systemic atrioventricular valve replacement/valvuloplasty improved systemic ventricular function for ccTGA with an intact ventricular septum. Physiologic repair was not ideal for ccTGA with PS. Severe systemic atrioventricular valve regurgitation and systemic ventricular dysfunction were associated with suboptimal outcomes.

Early Results of Robotically Assisted Congenital Cardiac Surgery: Analysis of 242 Patients

Robotic surgery has become a safe alternative to traditional and video-endoscopic approaches in cardiac surgery especially as favorable results have been reported in mitral valve procedures and totally endoscopic coronary artery bypass operations. Despite this, the adoption of robotics in congenital cardiac surgery has remained limited. In this study, the authors from Turkey reviewed their 8 year single-center experience with robotic congenital cardiac operations [5].

A total of 242 patients, (87.6% adults) underwent robotically assisted congenital operations. The authors approached peripheral cannulation via right internal jugular vein and right femoral vein and right femoral artery. Cardiac arrest was achieved by delivery of isothermic blood cardioplegia solution. All patients underwent a right chest approach. Nine patients had associated anomalies and majority of them were of venous system. Most common defects repaired were a secundum type atrial septal defect (157, 64.9%) and partial anomalous pulmonary venous drainage (PAPVC) (22, 9.1%). Mean patient age was 30.9 + / − 12 years and only 12% of patients were less than 18 years old.

Operative complications included atrogenic aortic laceration after declamping ($n=1$), aortic puncture site bleeding ($n=1$), laceration of visceral pleura ($n=1$), laceration of the superior vena cava ($n=1$), injury to the left atrial appendage by suction tip and femoral artery laceration ($n=1$). There was no operative mortality. Ventilation time, intensive care unit stay, and the length of hospital stay were 5.2 + / − 2.9 h, 16.8 + / − 2.5 h, and 3.5 + / − 1.1 days, respectively. The follow-up period was a mean 3.6 years (range, 6 months to 7.2 years). There was no mortality. Only 1 (0.4%) patient underwent reoperation via median sternotomy due to patch dehiscence after primum ASD repair. The other patients were asymptomatic through the follow-up period. No residual shunting, systemic or pulmonary venous inflow obstruction following PAPVC repair, outflow tract stenosis, or untreatable arrhythmias were detected.

The authors concluded that robotic technology can be utilized to perform suitable congenital operations safely and effectively albeit there is a learning curve.

Table 1 Summary of the studies in this review

Author	Study summary
Dovjak et al.	Abnormal Extracardiac Development in Fetuses With Congenital Heart Disease. [1] Extracardiac anomalies (ECA) in fetuses with congenital heart disease (CHD) can inform prenatal and postnatal decision making 429 fetuses with CHD who had a fetal MRI between 17–38 weeks of gestation 243/429 (57%) with ECA and 109 (25%) with structural brain anomalies (SBA) The most common ECA after SBA were extrafetal (21%) followed by urogenital (11%) Among fetuses without genetic anomalies, the highest ECA rate was in ventricular/atrial septal defects and the lowest in transposition of great arteries (21%) Fetal MRI shows a high prevalence of ECA and SBA in fetuses with CHD. Fetal MRI may aid the accurate prenatal diagnosis of ECA in CHD and thus help prenatal counseling of families with CHD
Truong et al.	Clinically Suspected Myocarditis Temporally Related to COVID-19 Vaccination in Adolescents and Young Adults This study evaluated the prevalence and the clinical course of post COVID-19 vaccination myocarditis,30 days after the vaccination 139 adolescents and young adults with 140 episodes of suspected myocarditis The median age was 15. 8 years 91% male, 66% white and 21% Hispanic 94% occurred after the Pfizer-BioNTech vaccine and 91% occurred after the second dose Almost all with chest pain. All had elevated troponin, 70% had abnormal EKG and or arrhythmia and 19% had decrease ejection fraction by echocardiogram Among the 97 patients who had a cardiac MRI at a median of 5 days, 76% had late gadolinium enhancement, 54% had myocardial edema while 51% met the Lake Louise criteria for myocarditis All the patients who had decreased ejection fraction by echo had a follow up echocardiogram that showed normal function The study concluded that almost all patients with post vaccine myocarditis had relatively mild course with normalization of ventricular function. Future studies for long term outcomes are needed
Meyer et al.	Integrated Clinical and Magnetic Resonance Imaging Assessments Late After Fontan Operation This large study evaluated the integration of clinical and CMR variables to identify patients with worse outcomes after the Fontan operation Retrospective study with 416 patients after the Fontan operation median age of 16 years. 57 patients (14%) with the composite outcome (46 death, 7 heart transplant and 4 heart transplant listing) Elevated ventricular end diastolic volume (EDVi) was the strongest risk factor for the composite outcome with a threshold above 156 ml/BSA$^{1.3}$ Ventricular dilation by CMR was the strongest predictor of worse outcomes along with other risk factors including worse circumferential strain, worse functional status, and worse global function index
Liu et al.	The Fate of Congenitally Corrected Transposition of the Great Arteries (ccTGA) Unoperated Before Adulthood [4] The outcomes, therapeutic strategies, and risk factors associated with ccTGA combined with associated cardiac lesions remain unclear 117 patients over a span of 9 years 3 groups: IVS, VSD and PS The PS groups had significantly less systemic AVV regurgitation and ventricular dysfunction and highest systemic ventricle ejection fraction 49 patients underwent surgery with no hospital mortality IVS group: patients receiving systemic atrioventricular valve replacement/ valvuloplasty had a significantly increased systemic ventricular ejection fraction and statistically more freedom from death and transplant than unoperated VSD group: late systemic ventricular ejection fraction of operated patients was not statistically different PS group: had significantly decreased systemic ventricular ejection fraction PS protects against systemic atrioventricular valve regurgitation and ventricular dysfunction Physiologic repair was not ideal for ccTGA with PS. Severe systemic atrioventricular valve regurgitation and systemic ventricular dysfunction were associated with suboptimal outcomes
Onan et al.	Early Results of Robotically Assisted Congenital Cardiac Surgery: Analysis of 242 Patients [5] 242 patients underwent robotic surgery from 2013–2020 Most common defect: secundum ASD, 74.7%; sinus venosus ASD 16%, PAPVC10.7% No mortality Operative complications included aortic laceration, aortic bleeding, pleural bleeding, injury to SVC, left atrial appendage and femoral artery Ventilation time, intensive care unit stay, and the length of hospital stay were 5.2 +/− 2.9 h, 16.8 +/− 2.5 h, and 3.5 +/− 1.1 days, respectively Postoperative rates of stroke, cardiac events, pulmonary complications, and reexploration were 0.4%, 2.4%, 4.1%, and 0.8%, respectively Robotic technology can be utilized to perform suitable congenital operations safely and effectively albeit there is a learning curve

Table 1 (continued)

Author	Study summary
Riggs et al.	Risk of Pediatric Cardiac Surgery Increased in Patients Undergoing Tracheal Surgery During the Same Hospitalization [6] Most common airway anomaly: congenital tracheal stenosis, tracheoesophageal fistula PHIS database, < 18 years old, 2005–2014 2 groups: CHS + TS, CHS alone propensity matched 2:1 Total number of patients 46,497, 283 in CHS + TS and 566 in CHS only group CHS + TS – increased mechanical ventilation, acute kidney injury, total parenteral nutrition Hospital mortality 13.8% for CHS + TS, 5.8% for CHS group Surgical complications 59% CHS + TS, 43% CHS group MV analysis risk factors for mortality: TS, ECMO, acute kidney injury, TPN The authors concluded that patients undergoing cardiac and TS in the same hospitalization are at greater risk of in-hospital mortality than patients undergoing similar cardiac surgeries alone and incur higher resource utilization thereafter. The increased risk of mortality is currently underappreciated, but it is important to recognize when discussing expectations with families and providers

ASD atrial septal defect, *AVV* atrioventricular valve, *CHS* congenital heart surgery, *ECMO* extracorporeal membranous oxygentation, *MV* multivariable, *TPN* total parenteral nuitrition, *TS* tracheal surgery, *IVS* intact ventricular septuam, *PAPVC* partial anomalous pulmonary venous return, *PS* pulmonary stenosis, *VSD* ventricular septal defect

Risk of Pediatric Cardiac Surgery Increased in Patients Undergoing Tracheal Surgery During the Same Hospitalization

The 2 most common airway anomalies requiring surgery in pediatric patients are congenital tracheal stenosis (CTS) and tracheoesophageal fistula (8). The outcome of surgical repair of isolated CTS is well documented. Operative survival ranges from 84 to 95% in larger series, although mortality as high as 30% is reported. The authors hypothesized that surgical repair of congenital tracheal anomalies and congenital heart disease (CHD) during the same hospitalization may still be associated with increased surgical risk [6].

Data were extracted from the PHIS database and comprised of all patients less than 18 years old from 2005 – 2014 who underwent cardiac operations. These were then divided into two groups: patients undergoing tracheal and cardiac surgeries (CHS + TS) during same hospitalization and those undergoing cardiac surgery alone (CHS). The groups were then propensity matched 2:1. A total of 46,497 patients underwent congenital heart operations. Of those, 283 patients with CHS + TS were matched to 566 patients with CHS only. CHS + TS was associated with more mechanical ventilation (98.2% vs 81.4%), acute kidney injury (15.5% vs 6.0%), and total parenteral nutrition (79.9% vs 46.3%), all $P < 0.001$, during their hospitalization.

The hospital mortality was 13.8% ($n = 39$) for CHS + TS patients and 5.8% ($n = 33$) for CHS only patients, P-value < 0.001. Surgical complication rates were 59.0% ($n = 167$) in CHS + TS patients compared to 43.3% ($n = 245$) in CHS only patients ($P < 0.001$). CHS + TS patients also had greater length of stay (63 vs 12 days). A multivariate analysis of the entire population found TS during the hospitalization to be a risk factor for mortality (hazard ratio [HR] 1.95, 95% CI 1.11–3.46, $P = 0.021$) as well as having extracorporeal membranous oxygenation (ECMO) during their stay (HR: 9.92, 95% CI 5.07–19.38, $P < 0.001$), experiencing acute kidney injury (HR: 4.25, 95% CI 2.26–8.00, $P < 0.001$), or requiring total parenteral nutrition (HR: 4.02, 95% CI 1.59–10.12, $P = 0.003$) (Table 1).

The authors concluded that patients undergoing cardiac and TS in the same hospitalization are at greater risk of in-hospital mortality than patients undergoing similar cardiac surgeries alone and incur higher resource utilization thereafter. The increased risk of mortality is currently underappreciated, but it is important to recognize when discussing expectations with families and providers.

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Declarations

Conflict of interest All authors declare that they have no conflict of interest.

Ethical approval Not applicable.

Consent to participate Not applicable.

Consent for publication Not applicable.

References

1. Dovjak GO, Zalewski T, Seidl-Mlczoch E, Ulm PA, Berger-Kulemann V, Weber M et al (2021) Abnormal extracardiac development in fetuses with congenital heart disease. J Am Coll Cardiol 78(23):2312–2322
2. Truong DT, Dionne A, Muniz JC, McHugh KE, Portman MA, Lambert LM et al (2021) Clinically suspected myocarditis temporally related to COVID-19 vaccination in adolescents and young adults. Circulation. https://doi.org/10.1161/CIRCULATIONAHA.121.056583
3. Meyer SL, St Clair N, Powell AJ, Geva T, Rathod RH (2021) Integrated clinical and magnetic resonance imaging assessments late after fontan operation. J Am Coll Cardiol 77(20):2480–2489
4. Liu R, Pang K, Li S, Zhang B, Rui L, Lin Y et al (2021) The fate of congenitally corrected transposition of the great arteries unoperated before adulthood. Ann Thorac Surg 112(6):2029–2037
5. Onan B, Onan IS (2021) Early results of robotically assisted congenital cardiac surgery: analysis of 242 patients. Ann Thorac Surg 112(6):2020–2027
6. Riggs KW, Wells D, Zafar F, Morales DLS, Bryant R, Tweddell JS (2021) Risk of pediatric cardiac surgery increased in patients undergoing tracheal surgery during the same hospitalization. World J Pediatr Congenit Heart Surg 12(6):730–736