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### INTRODUCTION

Like in the previous issue, we will present a table [Table 1] that summarizes significant publications in pediatric cardiac sciences that have appeared in the recent months. We have selected two papers for a detailed review. In the forthcoming issues we will seek to improve this section further. We would especially solicit contributions from trainees and fellows who could chose a recent publication in any of the pediatric cardiac specialties — cardiology, cardiac surgery, intensive care, and anesthesiology. Papers selected for journal clubs in their respective institutions can be discussed at length. It is particularly useful if the discussions include a critique as well as an effort to contextualize the paper to the low resource environments.

Olsson KM, Delcroix M, Ghofrani HA, Tiede H, Huscher D, Speich R, *et al.* Anticoagulation and survival in Pulmonary Arterial Hypertension: Results from the COMPERA Registry: Circulation 2014;129:57-65.

Pulmonary arterial hypertension (PAH) represents a heterogeneous group of disease entities. A vast number of conditions eventually culminate in increased pulmonary vascular resistance (PVR). The prognosis is, to some extent, influenced by the specific underlying cause of PAH. Wagenvoort and Wagenvoort examined lung biopsy samples of 156 patients with idiopathic (then termed primary) pulmonary hypertension and identified thrombosis of the small pulmonary vessels in a large number of patients.<sup>[1]</sup> He suggested that chronic thrombosis might contribute to the pathogenesis of this condition. Interest in therapeutic anticoagulation for patients with pulmonary hypertension stepped up after his paper and a number of descriptive studies suggested a survival benefit for patients with pulmonary hypertension on anticoagulants. However, no randomized control trials have been conducted and the

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ancillary nature of such treatment will make conducting such studies very difficult.

The Comparative, Prospective Registry of Newly Initiated Therapies for Pulmonary Hypertension (COMPERA) is a registry of pulmonary hypertension patients from 41 centers in seven European countries which records data regarding the demographics, severity, functional class (FC), homodynamic, and pharmacological information on newly diagnosed patients with PAH and follows them prospectively for a set of predefined significant clinical events. The study authors analyzed the database for outcome of newly diagnosed PAH patients from 2007 to 2013 with and without anticoagulation as well as subset analysis of idiopathic PAH (IPAH) and PAH due to other causes. The attrition rate on follow-up in the registry was less than 2%.

There were 1,283 newly diagnosed patients with PAH in the registry. Fifty-eight percent of these patients received anticoagulation (90% were on warfarin and the remaining were on heparin or novel oral anticoagulants). The target international normalized ratio (INR) at all participating centers was between 2 and 3. The duration of anticoagulation during the observation period varied among the patients. 16.4% of patients in the anticoagulation group and 18% of patients in the non-anticoagulation group died during the observation period. Subgroup analysis of IPAH and familial PAH revealed that 66% of 800 patients received anticoagulation. Fourteen percent of IPAH patients who received anticoagulation and 21% of patients who did not, died during the observation period. When the baseline hemodynamics was analyzed, it was noticed that the patients on anticoagulation represented a more severe subset of the disease population. Hence, the study group matched 168 patients in each group on the basis of age, sex, function class, and baseline PVR and demonstrated a statistically significant survival advantage for patients on anticoagulation. On multivariate analysis, male patients, older age group, and patients in FC IV were found to have an increased risk for death, while anticoagulation therapy was the only factor noticed to be protective. These patients were however not matched on the basis of treatment and a larger number of patients in the anticoagulation group were on combination therapy. Four of the 75 deaths in the anticoagulation group were

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# Table 1: A summary of significant publications in pediatric cardiac sciences in periodJune 2014-November 2014

Category	Reference	Study design	Study question	Conclusion
Pulmonary hypertension	Anticoagulation and survival in pulmonary arterial hypertension: results from the Comparative, Prospective Registry of Newly Initiated Therapies for Pulmonary Hypertension (COMPERA). Olsson KM, Delcroix M, Ghofrani HA, Tiede H, Huscher D, Speich R, Grünig <i>et al.</i> Circulation 2014;129:57-65	Prospective registry: Comparative, Prospective Registry of Newly Initiated Therapies for Pulmonary Hypertension (COMPERA)	Whether anticoagulation improves survival in idiopathic Pulmonary arterial hypertension (PAH)? Can anticoagulation benefit other forms of PAH also?	In a cohort of 1,283 consecutively enrolled newly diagnosed PAH, anticoagulation was used in 66% of 800 patients with IPAH and in 43% of 483 patients with other forms of PAH. In patients with IPAH, there was a significantly better 3-year survival. The evidence for benefit of anticoagulation for other forms of PAH not strong
Pediatric cardiac intervention	Palliative stenting of patent ductus arteriosus in older children and young adults with congenital cyanotic heart disease. Francis E, Kumar S, Kumar RK. Catheter Cardiovasc Interv. 2014:83:1109-15.	Retrospective	Whether patent ductus arteriosus (PDA) stenting is effective palliation in older children (>2 years)	Fifteen patients (age median: 14 years, range: 2-18 years) had palliative PDA stenting. All had improved saturation without any major adverse effect, making it a viable option in selected older patients with congenital heart disease and critically reduced pulmonary blood flow
General pediatric cardiology, arrhythmia, and electrophysiology	Usefulness of Arrhythmias as Predictors of Death and Resource Utilization in Children With Myocarditis. Anderson BR, Silver ES, Richmond ME, Liberman L. Am J Cardiol. 2014 Aug 12. pii: S0002- 9149:01595-1.	Retrospective cohort	Whether pediatric myocarditis patients with tachyarrhythmia have poor outcome?	Retrospective multicentric US evaluation of 2,041 myocarditis patients over 10 years showed incidence of tachyarrhythmia 11.5% and bradyarrhythmia 1.1%. The overall mortality was 8.7%. Tachyarrhythmia was 2.3 times associated with mortality. It was also associated with longer bosnital stav
General pediatric cardiology	β-Blockers and Angiotensin Converting Enzyme Inhibitors: Comparison of Effects on Aortic Growth in Pediatric Patients with Marfan Syndrome. Phomakay V, Huett WG, Gossett JM, Tang X, Bornemeier RA, Collins RT. J Pediatr. 2014 Aug 7. pii: S0022-3476-00630-1	Retrospective cohort	Comparing efficacy of beta blocker and angiotensin converting enzyme inhibitors (ACEI) in reducing aortic growth velocity (AGV) in Marfan Syndrome (MFS)	Sixty-seven MFS patients were retrospectively analyzed with follow-up for $7 \pm 6$ years. B-blocker therapy was found causing near-normalization of AGV in MFS. ACEI did not decrease AGV in a clinically significant manner
Anesthesia	The role of plasma Gelsolin in cardiopulmonary bypass induced acute lung injury in infants and young children: A pilot study. Shi S, Chen C, Zhao D, Liu X, Cheng B, Wu S, Lin R, Tan L, Fang X, Shu Q. BMC Anesthesiol. 2014;14:67.	Prospective analysis of plasma gelsolin level in 77 patients (<3 years) undergoing cardiac surgery	Role of gelsolin in predicting acute lung injury (ALI) after cardiopulmonary bypass (CPB)	Patients developing CPB-ALI had lower plasma gelsolin reservoir and a much more amount and rapid consumption of plasma gelsolin (PGSN) early after operation. PGSN before CPB was an early and sensitive predictor of CPB-ALI in infants and young children undergoing cardiac surgery, and was negatively correlated with the severity of CPB-ALI
General pediatric cardiology, arrhythmia, and electrophysiology	Flecainide Use in Children with Cardiomyopathy or Structural Heart Disease. Moffett BS, Valdes SO, Lupo PJ, delaUz C, Miyake C, Krenek M, Kim JJ. Pediatr Cardiol 2014 Aug 9.	Retrospective cohort	How safe is flecainide use in children with coronary heart disease (CHD) or cardiomyopathy (CM) and its association with cardiac arrest or death	42 hospitals collective data of antiarrhythmic drug use for CHD/CM pooled. Flecainide was administered in 229 patients. 6.5% in a cohort of 3,544 patients with CHD or CM (median age 73 days). The incidence of cardiac arrest in patients with CHD or CM receiving flecainide was 3.0% with an overall mortality of 4.3%.
Pediatric cardiac surgery, catheter intervention	Incidence and outcomes of right- sided endocarditis in patients with congenital heart disease after surgical or transcatheter pulmonary valve implantation. Malekzadeh-Milani S, Ladouceur M, Iserin L, Bonnet D, Boudjemline Y. J Thorac Cardiovasc Surg 2014	Retrospective cohort study	Analysis and outcome comparison of all right-sided endocarditis in surgical/ percutaneous pulmonary valve implant (PPVI)	There were 31 cases with right-sided endocarditis post pulmonary valve implant, incidence of IR higher in PPVI (1.2 vs 3.9/100 patients). The clinical presentation, microbiology, and outcome were similar except that time of development of IE was earlier in PPVI. The role of bovine jugular veins in the development of endocarditis is concerning. However, despite a higher incidence of endocarditis in the PPVI group, the probabilities of survival and event-free survival were similar to the

surgical group

# Table 1: (Continued)

Category	Reference	Study design	Study question	Conclusion
Pediatric cardiac surgery	Prophylactic peritoneal dialysis catheter does not decrease time to achieve a negative fluid balance after the Norwood procedure: A randomized controlled trial. Ryerson LM, Mackie AS, Atallah J, Joffe AR, Rebeyka IM, Ross DB, Adatia I. J Thorac Cardiovasc Surg. 2014.	Prospective randomized trial	Whether prophylactic peritoneal dialysis catheter (PDC) reduces time to achieve negative fluid balance in Norwood patients?	In a cohort of 22 Norwood patients, 10 randomized for peritoneal dialysis and 12 standard therapy. Mean time for negative fluid balance comparable in both group (~2 days) and there was no difference between the two groups lactate reduction, inotrope score, time to sternal closure, time to first extubation, and hospital length of stay. Additionally there were adverse events in PDC group. The PDC was not found superior to standard therapy at cost of adverse events
Imaging/ echocardiography	Prognostic implications of the systolic to diastolic duration ratio in children with idiopathic or familial dilated cardiomyopathy. Mondal T, Slorach C, Manlhiot C, Hui W, Kantor PF, McCrindle BW, Mertens L, Friedberg MK.Circ Cardiovasc Imaging 2014 <sup>-7</sup> :773-80	Retrospective cohort study, serial assessment of echocardiographic parameter for prognostication	Whether systolic to diastolic time duration ratio (S/D) is a useful marker in prognosis of mortality/ transplant in children with dilated cardiomyopathy (DCM)?	Forty-eight patients (~7 years) with DCM had serial evaluation of S/D ratio with 25-matched control. A systole to diastole duration ratio of >1:2 was found significantly linked with mortality and transplant
Pediatric cardiac surgery	Management of an Associated Ventricular Septal Defect at the Time of Coarctation Repair. Plunkett MD, Harvey BA, Kochilas LK, Menk JS, St Louis JD. Ann Thorac Surg 2014.	Review of data submitted to the Pediatric Cardiac Care Consortium of patients undergoing repair of coenzyme A (CoA) from 1982 to 2007	What are the comparative outcomes for the strategies for repair of ventricular septal defect (VSD) with coarctation?	The association of CoA and VSD is common. A strategy of concomitant VSD closure at CoA repair does not result in worse discharge mortality when compared with pulmonary banding with anticipated staged repair of the VSD. These outcomes support continued evaluation of a one-stage approach
Pediatric cardiology, fetal catheter intervention	Fetal aortic valvuloplasty for evolving hypoplastic left heart syndrome: Postnatal outcomes of the first 100 patients.Freud LR, McElhinney DB, Marshall AC, Marx GR, Friedman KG, Del Nido PJ, Emani SM, Lafranchi T, Silva V, Wilkins-Haug LE, Benson CB, Lock JE, Tworetzky W2. Circulation. 2014;130:638-45.	Retrospective analysis of 100 fetuses undergoing mid gestation balloon aortic valvuloplasty(BAV)	What is the short and intermediate term outcome of fetal BAV?	Forty percent of the fetal BAV achieved biventricular circulation. The left-sided structures were better in the biventricular group and had minimal reintervention. Still 60% had univentricular path with less favorable outcome. Short- and intermediate-term survival among patients who underwent fetal aortic valvuloplasty and achieved a BV circulation postnatally is encouraging. However, morbidity still exists, and ongoing assessment is warranted
Pediatric cardiac surgery	Pulmonary Atresia/Intact Ventricular Septum: Influence of Coronary Anatomy on Single- Ventricle Outcome. Cheung EW, Richmond ME, Turner ME, Bacha EA, Torres AJ. Ann Thorac Surg 2014.	Retrospective analysis of pulmonary atresia intact ventricular septum patients undergoing single ventricle palliation	Role of coronary anatomy (RV dependent coronary circulation) in single ventricle palliation outcome	In patients with PA-IVS undergoing single-ventricle palliation, RVDCC is associated with high early mortality, especially with coronary ostial atresia. There should be early consideration of transplantation in neonates with RVDCC. Patients with non-RVDCC undergoing single-ventricle palliation have excellent long-term outcomes, with no mortality
Pediatric cardiac intervention (experimental)	A novel design biodegradable stent for use in congenital heart disease: Mid term results in rabbit descending aorta. Veeram Reddy SR, Welch TR, Wang J, Richardson JA, Forbess JM, Riegel M, Nugent AW. Catheter Cardiovasc Interv 2014;26.	Prospective experimental animal (rabbit) study	Feasibility of biodegradable poly L-lactic acid stent in CoA	Stent deployment inrabbit descending aorta showed good stent position at 9-month follow-up. There were some concern raised for potential embolized fragment hazard
Pediatric cardiac surgery, general pediatric cardiology	Evaluation of electrocardiographic changes after arterial switch operation. Amoozgar H, Amirghofran AA, Salaminia S, Cheriki S, Borzoee M, Ajami G, Peiravian F. Int Cardiovasc Res J. 2014;8: 99-104.	Case-control; 33 patients compared with 33 age and gender matched controls	What are the electrocardiographic changes in children who have previously undergone an arterial switch operation (ASO)?	In a cohort of 33 patients, the abnormalities included right bundle branch block (18 vs 0%), bifascicular (3 vs 0%), and first-degree blocks (15 vs 0%). The increased P dispersion in ASO patients may increase the risk of future atrial arrhythmia

# Table 1: (Continued)

Category	Reference	Study design	Study question	Conclusion
General pediatric cardiology	Beta-Blockers (Carvedilol) in Children with Systemic Ventricle Systolic Dysfunction — Systematic Review and Meta- Analysis. Prijic S, Kosutic J, Vukomanovic V, Prijic A, Bjelakovic B, Zdravkovic M. Rev Recent Clin Trials 2014;8.	Meta-analysis	Whether carvedilol is beneficial in systemic ventricle systolic dysfunction?	Eight prospective studies found and analyzed. Carvedilol was found beneficial in systemic ventricle systolic dysfunction. Odds ratio for chronic heart failure related mortality/ heart transplantation secondary to carvedilol was 0.57 (95% confidence interval (CI): 0.33-0.97). The analysis showed that carvedilol could prevent one death/heart transplantation by treating 17 pediatric patients with impaired systemic ventricle systolic function
Pediatric cardiac surgery, intensive care	Utility of Clinical Biomarkers to Predict Central Line Associated Bloodstream Infections After Congenital Heart Surgery. Shin AY, Jin B, Hao S, Hu Z, Sutherland S, McCammond A, Axelrod D, Sharek P, Roth SJ, Ling XB. Pediatr Infect Dis J 2014;17.	Retrospective comparative	What is the utility of commonly used biomarkers in predicting the presence of catheter associated blood stream infections after congenital heart surgery?	450 cases cohort, infection found in 27 cases that was compared with 270 noninfected cases. Younger age and complex heart surgery linked with infection. Though fever and elevated C-reactive protein (CRP) found in infection, but strength to link was not powerful
Catheter interventions	Outcomes Using a Clinical Practice Pathway for the Management of Pulse Loss Following Pediatric Cardiac Catheterization. Glatz AC, Keashen R, Chang J, Balsama LA, Dori Y, Gillespie MJ, Giglia TM, Raffini L, Rome JJ. Catheter Cardiovasc Interv 2014;26.	Prospectively collected quality improvement database, reviewed retrospectively	What is the best strategy to deal with pulse loss after pediatric cardiac catheterization?	In a cohort of ~1,674 patients, pulse loss was seen in 5.4%. Eight patients had persistent thrombus despite a full treatment course (89% success rate in those able to complete treatment). A protocol for managing post- catheterization pulse loss is incorporated in the paper
Pulmonary hypertension, imaging/ echocardiography	Tricuspid Annular Plane Systolic Excursion (TAPSE) Is Reduced in Infants with Pulmonary Hypertension. Zakaria D, Sachdeva R, Gossett JM, Tang X, O'Connor MJ. Echocardiography. 2014;7	Retrospective case-control	What is the role of TAPSE — tricuspid annular plane systolic excursion — in infants with PH and compares it with other markers	Thirty patients with pulmonary hypertension compared with 69 controls. The TAPSE was indexed to body surface area. They found TAPSE and TAPSE/indexed are reproducible marker of right ventricular (RV) systolic function in infants with PH and is superior to tricuspid annular S' and RV fractional area change
Fetal cardiology, general pediatric cardiology	Anti-Ro/SSA-p200 antibodies in the prediction of congenital heart block. An Italian multicenter cross-sectional study on behalf of the 'Forum Interdisciplinare per la Ricerca nelle Malattie Autoimmuni (FIRMA) Group' Scarsi M, Radice A, Pregnolato F, Ramoni V, Grava C, Bianchi L, Gerosa M, Mosca M, Ghirardello A, Tani C, Motta M, Quinzanini M, Tincani A, Ruffatti A, Migliorini P, Doria A, Meroni PL, Brucato A. Clin Exp Rheumatol. 2014;20.	Retrospective; nested case- control in a subset	What is the association between the presence of specific anti-52 Ro/ SSA-p200 antibodies and congenital heart block?	Out of 207 pregnant women carrying anti-Ro/SSA antibody (Ab) analyzed, 42 children developed congenital heart block. Anti-p200 Ab were more frequently positive (81.0 vs 59.1%, P = 0.013) and at a higher titer in CHB mothers with an odds ratio of 2.98. This association was maintained even when the 42 mothers of children with CHB were compared with a control group matched for age and diagnosis (80.9 vs 50.0%; $P=0.006$ ). Anti-p200 antibodies seem to be associated with CHB with a higher probability than anti-Ro/SSA Ab, and therefore may be an additional test to identify mothers at higher risk to deliver affected children. An enzyme-linked immunosorbent assay (ELISA) screening for anti-Ro/SSA 60 kD Ab is nevertheless mandatory, given the probability of developing CHB also in the absence of anti-p200 Ab

directly attributed to a major bleeding event. There were four other admissions during the observation period for a nonfatal bleeding event. No data on minor bleeds were available in the registry. Analysis of the non-IPAH patients in the registry did not demonstrate a similar survival benefit for anticoagulation (21 vs 17% deaths) with a negative survival benefit noticed among the small subset of patients with scleroderma associated PAH.

The strengths of the study include the large number of patients, longer period of observation, and the low attrition rate. However, it is an observational study on the data from the registry and not a randomized controlled trial. The treatment strategy for a particular patient was based on the discretion of the treating physician. A larger number of patients in the anticoagulation group were on combination therapy that would have contributed to an improved control of pulmonary artery pressure. The registry data did not include minor bleeding events in patients on anticoagulation as well as information on the therapeutic range of anticoagulation in individual patients. Patients on anticoagulation were found to have more severe disease at diagnosis and, despite the abovementioned limitations, survived longer than patients without anticoagulation. This data cannot perhaps be ignored. Deaths directly attributable to a clinically significant bleeding event were infrequent during the observation period.

As the authors rightly pointed out in the discussion, most therapeutic trials on PAH (including newer pulmonary vasodilators and combination therapy) have been of shorter durations and have focused on functional endpoints of doubtful clinical significance (such as a small increase in 6-min walk test distance). Given the heterogeneity of the condition and the small number of patients in each subcategory, such registries will continue to be an important source of information for the management patients with PAH. Based on the evidence from this paper, it seems reasonable to consider anticoagulation in patients with IPAH and familial PAH.

Cheung EW, Richmond ME, Turner ME, Bacha EA and Torres AJ. Pulmonary Atresia/Intact Ventricular Septum: Influence of Coronary Anatomy on Single-Ventricle Outcome Annals of Thoracic Surgery 2014;98:1371-7.

Pulmonary atresia with intact ventricular septum (PA-IVS) represents approximately 1% of all congenital heart diseases and is iconic of the heterogeneity that exists within most congenital cardiac defects. The morphological spectrum varies from a normal sized or dilated right ventricle (RV) to an extremely hypoplastic and hypertensive RV (the so called uniparitate RV) and diminutive tricuspid valve. The low incidence combined with the numerous morphological possibilities means that evidence-based guidelines for management does

not exist and the treatment has to be individualized to the particular patient's anatomy. RV to coronary artery fistulae have been reported in 30-68% of patients with PA-IVS and there is speculation that persistence of these fistulous communications may be the primary abnormality leading to the development of pulmonary atresia.<sup>[2]</sup> The extreme scenario is a RV-dependent coronary circulation (RVDCC) with little or no connection between the proximal coronaries and the aortic root.

Literature on the impact of a RVDCC on outcome of patients with PA-IVS is inconsistent. Early single institutional data suggested that RVDCC was significantly associated with poor outcomes.<sup>[3]</sup> The same conclusions were not replicated in the multi-institutional Congenital Heart Surgeons Society (CHSS) study in 2004 and a subsequent multicenter study in the United Kingdom.<sup>[4,5]</sup> However such multicenter studies, while providing data on a large number of patients, are challenged by inconsistencies in the definition of RVDCC between participating centers. The coronary vessels are frequently very small making delineation difficult even by angiography.

The paper that is being discussed here is a single institution study that retrospectively analyzed all patients with PA-IVS from 2000 to 2012. Thirty percent (17 of 58 children) of PA-IVS patients were considered for single ventricle palliation. All these patients had significant right ventricular hypoplasia. Eighty-eight percent of such patients had fistulous coronary communications of which 10 represented a RVDCC. In contrast, only 20% of biventricular repair patients had fistulous coronary connections. There were a total of five deaths and a single cardiac transplantation in the RVDCC group (60%). All deaths occurred within 6 months of age and all patients who had coronary ostial atresia died. Three of the deaths occurred after cardiac catheterization while awaiting a systemic to pulmonary artery shunt, while two events (one death and one transplant) occurred in the postoperative period. The last patient died suddenly at home at 6 months of age while awaiting superior cavopulmonary anastomosis. All patients had evidence of significant coronary insufficiency (electrocardiogram (ECG) changes, elevated troponin levels or autopsy evidence of coronary stenosis, and significant myocardial ischemia). The patients, who survived, underwent superior cavopulmonary anastomosis and three of them proceeded for Fontan completion successfully. Two of the three patients who underwent Fontan completion had evidence of myocardial ischemia on myocardial viability studies during follow-up. There were no deaths in the non-RVDCC group with one patient lost to followup. Repeat angiograms were available in seven patients. There were no changes in coronary artery lesions in the RVDCC group, while three of the four patients in the non-RVDCC group demonstrated significant reduction in the number and size of coronary artery fistulae.

The study impresses upon us the importance of defining the coronary anatomy comprehensively in all patients with PA-IVS before planning surgical repair. A combination of RV, aortic root, and selective coronary angiograms may be required to obtain good anatomic detail. Three of the deaths had a temporal association with cardiac catheterization and these patients require careful intra- and post-procedure monitoring preferably in an intensive care setting. There is a high interstage mortality in patients palliated along the single ventricle pathway and most of them may be related to coronary insufficiency. Some centers currently recommend primary cardiac transplantation for patients with very high-risk coronary anatomy (double coronary atresia).

Of greater interest, most of the patients had evidence of myocardial ischemia even after Fontan completion. PA-IVS probably represents a high-risk subset among patients with a Fontan circulation and requires closer monitoring for subclinical coronary insufficiency on follow-up. No management guidelines currently exist on management of such complications on follow-up. The fate of the fistulous ventricle to coronary artery communications is also gaining greater focus. While no changes were noticed in the RVDCC group, there was significant decrease in the burden of lesions in the non-RVDCC group. This is similar to data from other centers. Long-term follow up has shown two ventricle repair to be superior to Fontan palliation in PA-IVS and combined with promising results of ligation of the larger fistulae and growth of the RV after decompression, it would be worthwhile to aggressively pursue RV decompression in patients who have minor degrees of RV-coronary fistula and a reasonably favorable RV anatomy.

The obvious limitations of this retrospective study are the small number of patients and the complexity of cardiac morphology that greatly reduces the power of the study to identify risk factors. In limited resource environments, few centers currently operate on infants with complex cyanotic congenital heart diseases. It may be possible to define uniform case selection criteria for single versus two-ventricle repair and maintain a disease registry to evaluate outcomes. Such an exercise may prove worthwhile in improving our understanding of the disease and we may all be enriched by the experience.

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