Journal of the American College of Cardiology
© 2010 by the American College of Cardiology Foundation
Published by Elsevier Inc.

Vol. 55, No. 2, 2010
ISSN 0735-1097/10/$36.00
doi:10.1016/j.jacc.2009.08.040

YEAR IN CARDIOLOGY SERIES

The Year in Congenital Heart Disease

Thomas P. Graham, Jr, MD
Nashville, Tennessee

With the increasing number of excellent papers related to congenital heart disease, it has become more difficult to select those that are perceived to be particularly critical to scientific developments in the field. I have chosen in this review to focus almost entirely on results and to follow surgical- or catheter-based interventions that highlight the complex conditions that surgeons and cardiologists now are able to repair or palliate successfully. Sections chosen are tetralogy of Fallot (TOF), Fontan operation, cardiac surgery in general, transposition of the great arteries (TGA), hypoplastic left heart syndrome (HLHS), aortic valve surgery, aortoplasty, and ventricular septal defect (VSD) management. These particular areas were chosen because of the importance to the general field of congenital heart disease, and many of the areas highlighted continue to plague investigators as they attempt to improve long-term outcomes.

TOF

Papers about TOF include issues related to initial intervention assessment of pulmonary regurgitation (PR) timing of pulmonary valve replacement (PVR) as well as late follow-up and implication for management.

Tamesberger et al. (1) reported 90 consecutive infants who underwent early repair: 25 neonates repaired due to duct-dependent pulmonary circulation or severe hypoxemia and 65 infants <4 months old with elective repair. Their results indicated no 30-day mortality, and late mortality of 2% with median follow-up of 4.7 years. Seven of 88 (8%) needed reoperation and 12 of 88 required reintervention. The younger patients did not differ significantly in intensive care unit stay, ventilator days, hospital stay, complications, or reoperation. There were more frequent transannular patches and reinterventions in the younger infants. These are excellent results, but will such early repair result in more severe PR and the need for more frequent PVR?

Dohlen et al. (2) reported right ventricular outflow tract (VOT) stenting in 9 infants because of cyanosis or duct-dependency and adverse risk factors. Stenting improved O_2 saturation from 73% to 94%. Median left pulmonary artery (PA) *z*-score increased from −4.9 to −1.5, right PA increased from −3.7 to −0.8, and Nakata index increased from 56 to 150 mm^2/m^2. There were no procedural complications. Six patients have undergone repair with no deaths. This is another method to palliate infants who are duct-dependent with adverse risk factors and have a pulmonary valve that is deemed unsalvageable. Stenting allows infants to obtain a greater age and size when repair theoretically is safer.

Papadopoulos et al. (3) reported repair of pulmonary valves in 5 children and 2 adults. All had severe PR associated with right ventricular (RV) dilation and dysfunction after primary right VOT reconstruction. There were no operative or late deaths. All valves were repaired successfully with a mean PR grade of 1.3 post-operatively. Mean transvalvular gradient was 20 mm Hg for children and 23 mm Hg for adults. The mean RV dilation index decreased significantly from 0.85 to 0.6 for children and 1.4 to 0.9 for adults. No significant regurgitation of the reconstructed valve was observed in the 4-year follow-up. This may prove to be an alternative to PVR in selected patients.

van den Berg et al. (4) reported on biventricular (BV) reserve, N-terminal pro–B-type natriuretic peptide (NT-proBNP) levels, and exercise performance in relation to RV volume in 53 patients with PR after repair with a maximum age of 2 years at repair; interval since repair was 15 years with no residual lesions except PR. The mean RV end-diastolic volume was 140 ml/m^2. Median PR fraction was 37%. BV systolic stress response was normal, stroke volume increased, and no adverse effects to dobutamine infusion were encountered. NT-proBNP was increased in 2 patients, median level 10 pmol/l and levels correlated with PR percentage but not with RV size. Mean peak exercise O_2 consumption (VO_{2max}) was 96% of normal, and mean $workload_{max}$ was 89% of predicted. At midterm follow-up, NT-proBNP levels are normal and BV functional reserve and exercise tolerance are well preserved in TOF repaired at young age, irrespective of RV volume. This study questions the validity of isolated PR or RV volume criteria for PVR in this group. Low-dose dobutamine stress testing is well tolerated and may be a useful additional tool for decision making. These data indicate that age of repair may play a crucial role in the adaptation of the RV to volume load and may change the indications for PVR in those with early repair.

From the Division of Pediatric Cardiology, Vanderbilt Children's Hospital, Nashville, Tennessee.

Manuscript received July 1, 2009; revised manuscript received August 17, 2009, accepted August 24, 2009.

Knauth et al. (5) assessed ventricular size and function by cardiac magnetic resonance (CMR) as predictors of clinical outcomes after repair in 88 patients. The median time from repair to evaluation was 21 years, and median follow-up from baseline to most recent follow-up was 4 years. Adverse outcomes included death, sustained ventricular tachycardia (VT), and increase in New York Heart Association functional class to grade III or IV. Adverse outcomes occurred in 18 patients or 21%, death in 4, sustained VT in 8, and increase in New York Heart Association functional class in 10. Severe RV dilation and either left ventricular (LV) or RV dysfunction predicted major adverse clinical events. This information may help assist in terms of therapeutic interventions. It should be noted that age at repair and era of surgery also correlated with adverse outcomes although these effects were theoretically controlled for in the statistical models used in determining the effects of RV size and ventricular function.

Harrild et al. (6) studied 98 adults with late PVR for RV dilation. Matched controls were identified for 77 of these patients; controls had TOF with RV dilation and no PVR. Matching was done by age and baseline QRS duration. In the PVR group, 13 events occurred over 272 patient-years. In the matched comparison, no significant differences were seen in VT, death, or combined VT and/or death. This cohort experienced either VT or death every 20 patient-years, and in a matched comparison with a similar TOF group, late PVR for symptomatic PR/RV dilation did not reduce the incidence of VT or death. These results can help to guide expectations of patients and physicians alike; PVR can be performed too late to prevent a tragic outcome.

Godart et al. (7) used off-pump PVR with a self-expandable valve stent as an emerging technique in 15 lambs by direct access of the infundibulum through a left thoracotomy, combined with PA banding. Animals were followed by transesophageal echocardiography and after control hemodynamic study, were electively sacrificed (at day 7, month 1, or month 4). Two animals died after implantation (1 of pneumothorax and 1 of endocarditis). Doppler studies showed only mild PR. Systolic gradients were 18 mm Hg at 1 week, 13 mm Hg at 1 month, and 4 mm Hg at 4 months. Examination demonstrated connective tissue between the stent and pulmonary wall, which increased with time. This new hybrid approach using a porcine valve mounted in a self-expandable stent to replace the pulmonary valve, without cardiopulmonary bypass (CPB), can theoretically reduce morbidity and the potential for myocardial damage during open heart surgery.

Fontan Operation

Ongoing important studies continue to be related to post-Fontan natural history, methods of Fontan repair, flow dynamics, and evaluation before and after Fontan repair.

Shiraishi et al. (8) studied the impact of age at Fontan procedure completion on post-operative hemodynamics and exercise in 68 patients with a dominant LV. Patients underwent exercise testing and catheterization at 1 year and then every 5 years and were divided into Group A <3 years at repair and Group B ≥3 years. No patients died during follow-up of 91 months. Vo_{2max} was 61% of normal in Group A versus 52% of normal in Group B, and cardiac index was 3.3 versus 2.4 at 5 years and 3.3 versus 2.6 at 10 years. The LV ejection fraction (EF) at 10 years was 62 versus 53%, all higher in the younger group. Here is another vote for an earlier Fontan procedure to achieve a beneficial effect on exercise capacity and hemodynamics in patients with a dominant LV.

Kerendi et al. (9) reviewed 236 consecutive Fontan procedures; Group 1 (n = 21) had concomitant atrioventricular valve (AVV) repair or replacement as compared with Group 2 (n = 215) with no AVV surgery. AVV regurgitation was graded as 1 (none or trivial) to 4 (severe). Group 1 patients were older (4 years vs. 3 years) and had longer CPB times (118 min vs. 85 min) and aortic cross-clamp times (33 min vs. 14 min). There were no differences between groups in diagnosis, weight, hospital or intensive care unit stay, ventilator time, or 12-h chest tube output. Post-operative complications were similar between groups, including bleeding, neurologic injury, arrhythmias, and operative mortality. There was a significant decrease of AVV regurgitation after surgery (3 pre-operatively vs. 1.7 post-operatively). Successful AVV surgery can improve operative and late outcomes for single-ventricle patients. This study demonstrates that AVV surgery performed with the Fontan procedure increased operative times, but did not significantly increase morbidity or mortality and supports the use of AVV surgery for single-ventricle patients.

Ovroutski et al. (10) reported on 35 patients in whom Fontan operation was performed at a median age of 4.2 years and underwent angiographic measurements of the central and lower lobe PA diameter before and during a median follow-up of 4.6 years. Although somatic development was documented by body surface area measurements, the PA showed no gain in diameter, and PA and lower lobe indexes decreased significantly during follow-up. This phenomenon may lead to a gradual increase in pulmonary vascular impedance and could be a limiting factor for optimal Fontan circulation long term. This report emphasizes again the importance of PA size before operation.

Bacha et al. (11) reported on 49 patients who underwent connection of discontinuous PAs with or after a bidirectional Glenn (n = 29) or Fontan (n = 20) procedure at a median age of 7.9 years. The PA continuity was established by direct anastomosis in 27, interposition graft in 19, and transcatheter recanalization in 3. Survival was 92% at 1 year and 89% at 5 years. Recurrent PA occlusion was documented in 7 patients, 5 within 10 days of PA connection. The only factor associated with shorter freedom from PA occlusion was sole supply of blood flow to 1 lung by systemic-to-PA collaterals before connection. Among the 45 early survivors, freedom from PA reintervention or

occlusion was 83% at 1 year and 55% at 3 years. Discontinuous PAs can be successfully reconnected in most patients with a cavopulmonary connection, although nonconfluent PAs increase the risk of poor outcome after Fontan operation. The fact that recurrent PA occlusion was usually diagnosed early post-operatively indicates that early restudy is useful in all patients with less than optimal results. In patients with sole blood supply to 1 lung through collaterals, shunt placements before PA connection may optimize outcome.

Anderson et al. (12) from the Pediatric Heart Network Multicenter study reported on contemporary outcomes after the Fontan procedures in 546 children. Predominant ventricular morphology was 49% LV, 34% RV, and 17% mixed. Ejection fraction was normal for 73% of subjects; diastolic function grade was normal for 28%. Child Health Questionnaire scores were lower than for controls; however, over 80% of subjects were in the normal range. The BNP concentration ranged from −4 to 652 pg/ml. Mean percent Vo_{2max} was 65% and decreased with age. Ejection fracture and EF *z*-score were lowest and semilunar and AVV regurgitation were more prevalent in the RV subgroup. Older age at the time of the Fontan procedure was associated with more severe AVV regurgitation. Measures of ventricular systolic function and functional health status, although lower on average in the cohort compared with control subjects, were in the majority within 2 SDs of the mean for controls. The RV morphology was associated with poorer ventricular and valvular function. Effective strategies to preserve ventricular and valvular function, particularly for patients with RV morphology, are needed.

Kim et al. (13) reported on 114 patients who underwent superior vena cava-to-PA anastomosis as an adjunct to BV repair in patients with a questionably adequate pulmonary ventricle; median follow-up was 92 months. Patient groups were determined by small pulmonary ventricles, chronic pulmonary ventricle dysfunction facilitation of repair without ventricular dysfunction or functional problems of the pulmonary ventricles, and acute pulmonary ventricular dysfunction. The long-term outcome for operative survivors was 83%, 80%, and 69% at 5, 10, and 20 years respectively. Survival in the previous 10 years was 92%. Of the late deaths, 70% were known cardiac deaths or sudden. Patients with chronic pulmonary dysfunction demonstrated the best 10-year survival. Of the late survivals, 98% of patients are in New York Heart Association functional class I or II. Arterial oxygen saturation increased significantly from before to late after repair, 83% to 94%. Freedom from new atrial arrhythmia was 92% at 20 years. There was no patient with clinically evident protein losing enteropathy. The most common cause of late mortality was cardiac. Serious complication risk related to pulsatility in the superior vena cava was only 2.6%. This procedure usually relates to a small or dysfunctional RV and is an important adjunct for the 1.5 ventricle repair. Complications related to superior vena cava symptoms were rare and atrial defects usually closed spontaneously or by percutaneous occlusion.

Cardiac Surgery/General

These articles relate to congenital heart surgical outcomes for various diagnoses, complications of procedures, hybrid approaches, and practice patterns.

Polito et al. (14) reported the associations between intraoperative and early post-operative glucose levels and adverse outcomes after complex surgery for 378 consecutive high-risk patients. Intraoperatively, a minimum glucose ≤75 mg/dl was associated with greater adjusted odds of reaching the composite morbidity-mortality end point (odds ratio [OR]: 3.10). Greater duration of hyperglycemia (glucose: 126 mg/dl) during the 72 post-operative hours was associated with longer duration of hospitalization. In the 72 h after surgery, average glucose <110 or >143 mg/dl, minimum glucose ≤75 mg/dl, and peak glucose level ≥250 mg/dl were all associated with greater adjusted odds of reaching the composite morbidity-mortality end point. In children undergoing complex surgery, optimal, post-operative glucose range is probably 110 to 126 mg/dl and may require insulin infusions to achieve optimal outcomes. We usually think more about cardiac output, lactate levels, and low O_2 as indications for poor outcomes after surgery. This article showed that glucose levels may also be important and require tighter control than was previously practiced for optimal results.

Domi et al. (15) performed a case-control study of children from term birth to 18 years with congenital heart disease who underwent open heart surgery between January 1992 and March 2001 to document frequency, predictors, and neurologic outcomes of vaso-occlusive strokes. Case subjects experienced stroke within 72 h after surgery and control subjects (2 for each case subject) had surgery and no stroke. There were 30 children with stroke (28 with arterial ischemic stroke and 2 with cerebral sinovenosus thrombosis) who were identified among 5,526 children undergoing cardiac surgery yielding a risk for stroke of 5.4 per 1,000 children. Univariate analysis revealed that older age, longer duration of CPB, number of days in the hospital post-operatively, and reoperation were associated with stroke. In multivariate analyses, only reoperation was associated with stroke. Obviously, stroke is a devastating issue in children undergoing surgery, most of whom survive surgery and have a good cardiac outcome. In survivors, neurologic deficits were present in 72%, and 14% died after their stroke; only 14% had normal neurologic outcomes. Neurologic deficits include seizure disorder, headache disorder, as well as physical deficits. Patients with stroke were investigated for risk factors in addition to cardiac surgery. Prothrombotic abnormalities were found in 6% who tested positive for factor V Leiden mutation, 20% who tested positive for anticardiolipin antibody at least once, and 13% had a family history of thrombosis. Congenital and acquired prothrom-

JACC Vol. 55, No. 2, 2010
January 12, 2010:147–55

botic conditions may predispose patients to cerebral thrombosis and emboli. The study had insufficient power to identify predictors of stroke and potential preventative strategies. This is an important area of research and hopefully further studies can point to ways to decrease this devastating outcome of cardiac surgery.

Karamlou et al. (16) reported on practice patterns for management of adult congenital heart disease. These investigators identified patients with 12 congenital heart disease diagnostic groups using the Nationwide Inpatient Sample 1988 to 2003. Pediatric heart surgeons were identified as surgeons whose annual practice volumes were >75% pediatric heart cases. Operations on adults were identified within these 12 diagnoses occurring in patients ≥18 years of age. There were 30,250 operations yielding a national estimate of 152,277; 73% were pediatric operations and 27% were adult congenital heart disease operations. Pediatric heart surgeons performed 68% of pediatric operations in all diagnostic groups, whereas nonpediatric heart surgeons performed 95% of adult operations within the same groups. In-hospital mortality for adults operated on by pediatric heart surgeons were lower than that for patients operated on by nonpediatric heart surgeons: 1.87% versus 4.84%. Survival advantage increased with increasing annual pediatric volume.

Debate continues about how best to care for adult congenital heart patients. There are both pediatric and adult institutions that can show outstanding outcomes as long as they are equipped in terms of operating rooms, surgical and ancillary staff, and intensive care staff with training and experience to take care of them in either place. Familiarity with the type of complex anatomy one frequently is dealing with in these adult patients is critically important, and pediatric heart surgeons usually have this expertise to yield better outcomes.

Menon et al. (17) reported on hybrid intraoperative PA stent placement in 24 patients with a median age of 15 years (range 3 to 67 years). A total of 27 stents were deployed: left PA stent in 13, right PA stent in 8, and bilateral stents in 3 patients. Primary diagnoses were pulmonary atresia, TOF, tricuspid atresia, and other more complex conditions. Maximum balloon diameters ranged from 8 to 16 mm. Concomitant surgical procedures performed were RV to PA conduit replacement or right VOT reconstruction, PVR, and others. There were no deaths or PA damage. There were 2 cases of distal stent migration. Repeat stent dilations within 6 months were performed in 3 patients. Hybrid PA stenting can play an important role in the management of congenital heart disease with complex branch PA anatomy and can also be used as a rescue procedure following complications of percutaneous transcatheter procedures, such as stent embolization, which can be prevented by suturing the proximal end of the stent to the PA. There was no incidence of stent migration since this technique was instituted. The use of expandable stents may alleviate some of the questions about future intervention. Hybrid procedures are best carried out in a suite with biplane angiography available.

Vida et al. (18) studied patent ductus arteriosus ligation in premature babies with the question of optimal timing for surgical ligation. There were 201 premature babies (≤32 weeks gestation) from 2001 to 2007 who received primary medical treatment with ibuprofen. Medical treatment was effective in 149 patients, but 52 required surgical ligation after medical failure. Most who failed were younger, had lower weight, and had a higher incidence of symptomatic hypotension. More than 2 cycles of ibuprofen were associated with an increased risk for bronchopulmonary dysplasia (OR: 2.8) and acute renal failure (OR: 3.81). Prolonged patency of the ductus in pre-term infants is related to increased morbidity. Frequently, it is a difficult decision as to when to proceed with patent ductus arteriosus ligation in a premature infant, particularly in those who weigh 750 g or less. This study shows that more than 2 cycles of ibuprofen are not in the patient's best interest, and in this situation, patients should have their patent ductus arteriosus ligated.

Lim et al. (19) reported on the results in 2 patients with heterotaxy syndrome who underwent BV repair from 1990 to 2007. Left atrial isomerism was present in 73% and right atrial isomerism in 10%, with indeterminate atrial anatomy in 17%. Median age at repair was 6.8 months with a range of 5 days to 22 years. Systemic venous anomalies were present in 75 patients, pulmonary venous anomalies in 26, and endocardial cushion defects in 36. Operations included double switch repair, physiologic repair, arterial switch, and the Rastelli procedure. Separation of systemic from pulmonary venous return included intra-atrial baffle in 48 patients and extracardiac grafting in 2. Combined lesions were common, occurring in 99% of patients. Average follow-up was 45 months. Survival was 93% at 10 years; unbalanced atrioventricular canal was the only risk factor for mortality. Subsequent procedures were common with a 10-year freedom from reoperation or reintervention of 38%. Arrhythmias occurred in 40%, bradyarrhythmia in 30%, and tachyarrhythmia in 16% of patients. Freedom from any arrhythmia was 54% at 10 years. Despite complex anatomies, these patients should be better off in most cases with a BV versus Fontan operation when feasible.

TGA

Follow-up and natural history of patients undergoing atrial and arterial switch procedures are of considerable importance, as detailed in the following articles.

Hörer et al. (20) compared survival, freedom from reoperation, and functional status between atrial switch and arterial switch operations. There were 88 Mustard patients, 329 Senning procedures, and 512 arterial switch operations between 1974 and 2006. In-hospital mortalities were 8% for Mustard, 4.6% for Senning, and 6.4% for arterial switch.

Presence of VSD was the only risk factor for in-hospital mortality in multivariate analysis. Highest survival at 20 years was after arterial switch (96%), followed by Senning procedure (93%), and Mustard operation (82%). TGA with VSD, TGA with VSD plus left VOT obstruction, and Mustard operation emerged as risk factors for late death, with arterial switch as a protective factor. Highest freedom from reoperation at 20 years was after Senning procedure, followed by arterial switch and Mustard operation. Presence of complex transposition, previous palliative operation, surgery between 1985 and 1995, surgery after 1995, and Mustard operation emerged as risk factors for reoperation. Change from atrial to arterial switch led to improved long-term survival after hospital discharge but not to lower incidence of reoperation. This excellent documentation of a large number of patients followed before and after the change from atrial to arterial switch operation shows the suspected improvement in survival and functional status at this point 10 years out. It is interesting that the Senning patients do much better than Mustard patients do. This is probably because many Senning patients were operated on at a younger age and in an era of improved intraoperative myocardial protection. The fact that the arterial switch patients had a greater reoperation rate is due to the pulmonary stenosis problem early in the use of this operation as has been noted by other groups and has largely been circumvented by changes in the technique to reconstruct the pulmonary arteries.

Bové et al. (21) reported on 93 children who were reviewed for functional and morphologic assessment of both reconstructed arteries after the arterial switch. At a mean follow-up of 5 years, aortic regurgitation (AR) >2 developed in 10% in TGA with intact ventricular septum versus 23% in TGA with VSD. Neoaortic obstruction occurred in 8%, mostly at the neosinotubular anastomosis, and correlated with prior pulmonary to aortic ratio >1.5. Freedom from reintervention at 1, 5, and 10 years was 98%, 96%, and 96%, respectively, for TGA/intact ventricular septum versus 65%, 63%, and 63%, respectively, for TGA/VSD. An aortic arch obstruction and VSD were significant predictors for reintervention. After arterial switch operation, the neoaortic root is usually enlarged, but with a growth pattern comparable to that of a normal population. The presence of a VSD and larger aortic root size predisposes to both neoaortic valve dysfunction and root enlargement. Severe root dilation appears to be closely related to significant neoaortic valve regurgitation, as a result of a time-dependant and reciprocal process. Neopulmonary stenosis is a frequent finding, but rarely has clinical consequences. Significant AR reported here is of concern. This appears to be brought on mainly by the enlarged aorta at the time of operation usually associated with VSD and neoaortic obstruction. Although the PA appears to take on the characteristics of a normal aorta with time, particularly in those with younger age of operation, there still can be significant size discrepancy, which leads to ongoing AR. Hopefully, this will stabilize and not require a large number of patients requiring aortic valve surgery.

Petit et al. (22) reported follow-up after balloon atrial septostomy (BAS) for TGA. Twenty-six infants with TGA were retrospectively included from a larger cohort of infants with congenital heart disease who underwent pre-operative brain magnetic resonance imaging as part of 2 separate prospective studies. Data collected included all pre-operative pulse oximetry recordings, all values from pre-operative arterial blood gas measurements, and BAS procedure data. Magnetic resonance imaging scans were performed on the day of surgery, before the surgical repair. Of the 26 infants, 14 underwent BAS. No stroke was found in the entire cohort, whereas 10 of 26 patients were found to have hypoxic brain injury in the form of periventricular leukomalacia. This problem was not associated with BAS; however, neonates with periventricular leukomalacia had lower pre-operative oxygenation and a longer time to surgery than those without periventricular leukomalacia. Clearly, BAS was not associated with significant brain injury. Theoretically, BAS could cause brain injury with significant air embolus. However, this should be a rare complication.

Sharma et al. (23) reported on double switch operation or congenitally corrected TGA in 68 patients. Group 1 comprised 31 patients at a mean age of 94 months who underwent a combined Rastelli and atrial switch operation. Group 2 comprised 37 patients with a mean age of 36 months who underwent an arterial switch operation and atrial re-routing. Of the patients in Group 2, 8 had an intact ventricular septum. Group 1 had 5 early deaths (17%) but no late deaths. Three patients underwent conduit revision at a mean follow-up of 62 months. Group 2 had 5 early deaths (13.5%). There were 4 late reoperations (2 pulmonary baffle revisions, 1 mitral valve replacement, and 1 permanent pacemaker implantation) and 4 late deaths (1 secondary to pulmonary hypertension, 2 secondary to uncontrolled atrial tachyarrhythmia, and 1 secondary to pulmonary hypertension and right ventricular failure). In Group 2, 4 patients had a left ventricular ejection fraction <40%, 5 had moderate AR, 5 patients had symptomatic tricuspid incompetence, 1 had tricuspid stenosis, 1 patient had superior cava obstruction, and 3 patients received antiarrhythmic therapy. This is a complex procedure and unfortunately can be associated with multiple complications. One distressing feature of this follow-up is the discovery of significant LV dysfunction at midterm follow-up.

Scherptong et al. (24) studied the effects of tricuspid valve surgery in 16 adults with a dysfunctional systemic RV who underwent tricuspid valvuloplasty in 8 or replacements in 8. There were 9 patients with congenitally corrected TGA and 7 with previous atrially corrected TGA. The patient ages averaged 35 years. Tricuspid regurgitation was graded 1 to 4 according to its severity, RV dysfunction was graded as to 1 to 4, with 1 = no dysfunction to 4 = severe dysfunction. Although complications occurred in 11 patients, all could be

managed. Three patients died 109 to 659 days after surgery. Overall, tricuspid valve function improved from grade 3 to 0.9 and functional class improved from 2.7 to 2.1, whereas RV function remained unchanged. After surgery, however, recurrent moderate tricuspid regurgitation was observed frequently (37%). Although this study suggests you can buy a bit of time with target vessel revascularization in patients with a dysfunctional systemic RV, the results are not overwhelmingly positive. Certainly one should consider this possibility in patients who have only mild RV dysfunction. Another option that may be useful is PA banding in those with low LV pressure as this relatively benign type of surgery can result in improvement in tricuspid valve function and stabilization of patients. Unfortunately, rehabilitation of a systemic RV is fraught with difficulty in any type of surgical intervention in the adult and transplant may be the best option for many patients.

HLHS

Approaches to risk stratification in infants with HLHS and outcomes continue to be of importance.

Honjo et al. (25) reported on clinical outcomes, program evolution, and PA growth in single-ventricle palliation using hybrid and palliative strategies. In all, 58 patients underwent Norwood operation with Blalock-Taussig shunt (in 39) or hybrid procedures (in 19). At pre-stage 2 evaluation, there were nonsignificant trends toward lower ventricular end-diastolic pressure with higher mixed venous saturation, and larger Nakata and lower lobe indexes in the hybrids. Mean PA pressures were not different between the groups. Four Norwood patients (10%) underwent transplantation before stage 2 palliation. Forty-two patients underwent stage 2 palliation or stage 2 hybrid procedure. Requirement for PA-plasty, postoperative central venous pressure, stage 2 survival, and 1 year survival were similar between groups. Combined (stage 1 plus stage 2) intubation time, intensive care unit time, and hospital length of stay was shorter for hybrids than for Norwood survivors. Comparison of resource utilization at the time of arch reconstruction demonstrated a time-related trend toward improvement in the hybrid group but not in the Norwood group. Hybrid palliation does not have a significant adverse impact on PA development, with comparable PA growth and hemodynamics. The hybrid palliative strategy has emerged as an alternative to Norwood palliation. An intuitive but unproven advantage of this strategy is the avoidance of CPB and aortic arch reconstruction in the neonatal period and deferring these procedures until the stage 2 procedure at 4 to 6 months of age when the patient is hypothetically more able to tolerate a "big" operation. There are patients in whom this is not an acceptable procedure, particularly in those who have narrowing of the pre-ductal arch, which leads to severe acute coronary ischemia or after stent placement. It does appear that hybrid palliation in this study showed improved RV function as compared with RV function of the classical Norwood group. As noted in the discussion of this article, it may be time for a comparison of hybrid versus standard surgery for Norwood patients and in a large multi-institution study.

Tabbutt et al. (26) studied neurodevelopmental outcomes after staged palliation for HLHS at 1 year of age. Entry criteria included all patients with single-ventricle physiologic features and systemic outflow obstruction undergoing staged palliation. Exclusion criteria before surgical intervention included: 1) multiple congenital anomalies; 2) recognizable chromosomal or phenotypic syndrome at birth; and 3) non-English language primarily spoken in the home. A total of 83 patients underwent multiple operations with CBP during the first year of life. The mean was 2 operations, with 7 patients requiring extracorporeal membrane oxygenation palliation post-operatively. There were 25 patients or 28% with a confirmed or suspected genetic syndrome. At 1 year of age, the neuromuscular examination results were abnormal or suspect for 65%. The mean Mental Developmental Index score was 90, and 10 patients had scores <70. The median Psychomotor Development Index was 73, and 42 patients had scores <70. In multivariate analyses, younger gestational age, the presence of a genetic syndrome, the need for pre-operative intubation had significant negative effects on neurodevelopmental outcomes. No association was found with the operative factors, including duration of deep hypothermic circulatory arrest. The study suggests that future research should continue to focus on the role of perioperative management, post-discharge care, and early interventions. Longer-term developmental follow-up evaluation of these patients is of utmost importance and currently is underway. In addition, continued investigation to identify modifiable risk factors in this high-risk group of infants should be supported.

Grosse-Wortmann et al. (27) used CMR in 20 consecutive patients with the mean age of 10 days to assess LV volume in terms of decision-making of BV versus univentricular repair. Diagnoses were aortic stenosis in 3 patients, HLHS in 12 patients, and unbalanced atrioventricular septal defect in 5 patients. Potential LV volumes assuming an ideal geometric shape were calculated by mathematically "unfolding" the compressed LV. The left ventricular end-diastolic volume (LVEDV) was 16 ± 7.0 ml/m^2 by echocardiography and 33.5 ± 15.5 ml/m^2 by CMR. Echocardiography consistently underestimated LV volume and did not correlate with CMR. Of all echocardiographic parameters, mitral valve z-score was the best predictor of LVEDV by CMR. The average potential volume increase was 8.8% for aortic stenosis, 35% for atrioventricular septal defect, and 23% for HLHS complex patients; 16 of 20 patients underwent BV repair without mortality. Of these, only 5 had a pre-operative LVEDV of more than 20 ml/m^2 by echocardiography. These investigators have shown an ability to get borderline LV volume patients through a BV operative plan by using CMR to more adequately estimate their volume and calculate potential volume, which is significantly augmented particularly in atrioventricular septal defect and HLHS patients. It was of interest that mitral valve diameter and aortic annulus size were also

found to be larger by CMR than by echocardiography. This is an unusual finding; these measurements are usually quite accurate by echocardiography. These findings can hopefully be applied to other patients with borderline situations and correctly put them into a BV repair mode when feasible. It can obviously be disastrous to use the BV approach with patients whose LV is really untenable for BV repair. Thus, it is critically important to be accurate before proceeding with BV repair in borderline patients.

Aortic Valve Surgery and Aortopathy

Aortic valve and aortic root pathology continue to be prevalent issues to deal with in the congenital heart populations.

Alsoufi et al. (28) studied mechanical valves versus the Ross procedure for aortic valve replacement (AVR) in 346 children: 215 who underwent the Ross procedure and 131 who underwent AVR with a mechanical prosthesis. Patients undergoing the Ross procedure were younger, more likely to have a congenital cause, and less likely to have a rheumatic or connective tissue cause. They had a lower frequency of regurgitation, required more annular enlargement, and had less concomitant cardiac surgery. Competing-risk analysis showed that 16 years after AVR, 20% had died without subsequent AVR, 25% underwent a second AVR, and 55% remained alive without further surgery. After propensity adjustment, factors associated with early phase death included mechanical valves and a nonrheumatic cause. Freedom from homograft replacement after the Ross procedure was 82% at 16 years of follow-up. These outcomes are confounded somewhat from the standpoint of U.S. and European patients because of the significant number with rheumatic heart disease. Survival was excellent with the Ross procedure versus mechanical AVR, particularly in patients who had rheumatic fever. Despite the surgical complexity, the Ross procedure was associated with only a 2.3% mortality, which is similar to data from the International Registry. These investigators reported a high incidence of Ross failure in patients with active rheumatic fever at the time of AVR, patients with dilation of the left VOT >30 mm, and patients with an aortic annulus of 3 mm or more larger than the pulmonary annulus. They now offer the Ross procedure exclusively in patients <5 years of age. These data show excellent long-term follow-up data in patients with Ross procedure who are nonrheumatic in origin.

Cameron et al. (29) reported on aortic root replacement (ARR) in 372 Marfan patients with an evolution of repair over 30 years. These investigators report that of 269 of 372 patients with a Bentall composite graft, 85 had valve-spring ARR, 16 had ARR with homografts, and 2 had ARR with porcine xenografts. In the first 24 years of the study, 85% received a Bentall graft; during the last 8 years, 61% had a valve-sparing procedure. There was no operative or hospital mortality among the 327 patients who underwent elective repair; there were 2 deaths among the 45 patients who underwent emergent or urgent repair. There were 74 late deaths (70 Bentalls, 2 homografts, and 2 valve-sparing ARRs). The most frequent causes of late death were dissection or rupture of the residual aorta. Prophylactic surgical replacement of the ascending aorta in patients with Marfan syndrome has a low operative risk and can prevent aortic catastrophe in most patients. Valve-sparing procedures, particularly using the reimplantation technique with the Valsalva graft, show promise but have not yet proven as durable as the Bentall procedure. These investigators show excellent results in the evolution of this important procedure. The current guidelines are for aortic root replacement in Marfan syndrome with aortic sinus diameter 5 cm or greater (or 4.5 cm among patients with family history of rupture or dissection); ascending aortic dissection, whether acute or chronic; aneurysm growth more than 1 m/year; and worsening AR in a dilated root when a valve-sparing procedure is desired. The results are much worse in terms of late complications and mortality in those who have dissection before surgery, as one might predict.

Aalberts et al. (30) reported on the many faces of aggressive aortic pathology in Loeys-Dietz syndrome (LDS). LDS is a newly recognized disorder of connective tissue that shares overlapping features with Marfan syndrome and the vascular type of Ehlers-Danlos syndrome, including aortic root dilation and skin abnormalities. It can be recognized by craniofacial characteristics, such as hypertelorism, bifid uvula, or cleft palate in LDS type 1, whereas these are absent in LDS type 2. This is a very aggressive aortic pathology. These investigators found 9 LDS patients from 4 families, related their features to published cases, and discussed important aspects of the diagnosis and management of LDS in order to make clinicians aware of this syndrome. Because aortic dissection and rupture in LDS tend to occur at a younger age and because the vascular pathology can be seen throughout the entire arterial tree, patients should be carefully followed up and aggressive surgical treatment is mandatory. Pictures of patients in this article are quite useful in terms of discovering these patients and treating them in an aggressive fashion.

VSD Management

Yoshimura et al. (31) reported on surgical management of multiple VSDs using the felt sandwich surgical technique. There were 29 consecutive patients who underwent an operation for multiple VSDs and associated cardiac malformations including 17 male and 12 female infants with a median age of 6 months. Thirteen patients had 4 or more VSDs. There was no surgical or follow-up mortality, and no reoperations were required. There were no cases of heart block and no significant residual shunts in the latest follow-up study. Two patients with Swiss cheese septum had post-operative congestive heart failure; 3 muscular VSDs were closed with a sandwich technique in those patients. Of the other 27 patients, 7 of the 9 patients who

underwent the sandwich procedure had septal dysfunction, whereas 5 of the 20 other patients showed septal dysfunction. Although the sandwich technique is simple and effective, the use of numerous felt patches disturbed the movement of the interventricular septum. An effort should be made to close the muscular VSD directly to avoid post-operative cardiac dysfunction. Large apical defects, directly located underneath the moderator band, are considered suitable for the sandwich technique. Some of these defects can be closed with a percutaneous device; this may prove as useful as the felt sandwich technique and may result in less septal dysfunction, although this is unproven.

Chessa et al. (32) reported on 40 adult patients who underwent transcatheter closure of VSD. A shunt was considered significant for closure when left atrial or LV enlargement was found or there was a previous endocarditis. In 40 patients, 41 procedures were carried out; a muscular VSD occluder was used in 22 patients and a perimembranous VSD occluder in 18. No deaths occurred and no procedure was aborted. No device embolization occurred. The median follow-up duration was 36 months with a range of 6 to 81 months. One patient, who had 2 devices inserted because of a residual defect after TOF repair, had to be operated on again after the second device implantation because of a persistent residual leak. An internal jugular venous approach was used to close muscular VSDs whereas femoral vein access was used to close perimembranous VSDs. A total of 6 complications occurred. The most frequent complication was a rhythm abnormality directly after device implantation: 1 transient left anterior hemiblock, 1 transient complete AV block, and 2 ventricular fibrillations requiring electrical defibrillation; AR and/or tricuspid regurgitation are potential problems in these cases and were reported transiently in only 2 of 41 patients. This is a procedure for experienced interventionalists with surgical backup available at the time of the procedure.

Reprint requests and correspondence: Dr. Thomas P. Graham Jr., Vanderbilt Children's Hospital, Division of Pediatric Cardiology, 2200 Children's Way, Suite 5230, Nashville, Tennessee 37232-9119. E-mail: tom.graham@vanderbilt.edu.

REFERENCES

1. Tamesberger MI, Lechner E, Mair R, et al. Early primary repair of tetralogy of Fallot in neonates and infants less than four months of age. Ann Thorac Surg 2008;86:1928–36.
2. Dohlen G, Chaturvedi RR, Benson LN, et al. Stenting of the right ventricular outflow tract in the symptomatic infant with tetralogy of Fallot. Heart 2009;95:142–7.
3. Papadopoulos N, Esmaeili A, Zierer A, et al. Secondary repair of incompetent pulmonary valves. Ann Thorac Surg 2009;87:1879–84.
4. van den Berg J, Strengers JLM, Wielopolski PA, et al. Assessment of biventricular functional reserve and NT-proBNP levels in patients with RV volume overload after repair of tetralogy of Fallot at young age. Int J Cardiol 2009;133:364–70.
5. Knauth AL, Gauvreau K, Powell AJ, et al. Ventricular size and function assessed by cardiac MRI predict major adverse clinical outcomes and late after tetralogy of Fallot repair. Heart 2008:94:211–6.
6. Harrild DM, Berul CI, Cecchin F, et al. Pulmonary valve replacement in tetralogy of Fallot impact on survival and ventricular tachycardia. Circulation 2009;119:445–51.
7. Godart F, Bouzguenda I, Juthier F, et al. Experimental off-pump transventricular pulmonary valve replacement using a self-expandable valved stent: a new approach for pulmonary incompetence after repaired tetralogy of Fallot. J Thorac Cardiovasc Surg 2009;137: 1141–5.
8. Shirashi S, Yagihara T, Kagisaki K, et al. Impact of age at Fontan completion on postoperative hemodynamics and long-term aerobic exercise capacity in patients with dominant left ventricle. Ann Thorac Surg 2009;87:555–61.
9. Kerendi F, Kramer ZB, Mahle WT, et al. Perioperative risks and outcomes of atrioventricular valve surgery in conjunction with Fontan procedure. Ann Thorac Surg 2009;87:1484–9.
10. Ovroutski S, Ewert P, Alexi-Meskishvili V, et al. Absence of pulmonary artery growth after Fontan operation and its possible impact on late outcome. Ann Thorac Surg 2009;87:826–32.
11. Bacha EA, Lang P, Mayer JE Jr., McElhinney DB. Connection of discontinuous pulmonary arteries in patients with a superior or total cavopulmonary circulation. Ann Thorac Surg 2008;86:1948–54.
12. Anderson PAW, Sleeper LA, Mahoney L, et al. Contemporary outcomes after the Fontan procedure. A Pediatric Heart Network multicenter study. J Am Coll Cardiol 2008;52:85–98.
13. Kim S, Al-Radi O, Friedberg MK, et al. Superior vena cava to pulmonary artery anastomosis as an adjunct to biventricular repair: 38 year follow-up. Ann Thorac Surg 2009;87:1475–83.
14. Polito A, Thiagarajan RR, Laussen PC, et al. Association between intraoperative and early postoperative glucose levels and adverse outcomes after complex congenital heart surgery. Circulation 2008; 118:2235–42.
15. Domi T, Edgell DS, McCrindle BW, et al. Frequency, predictors, and neurologic outcomes of vaso-occlusive strokes associated with cardiac surgery in children. Pediatrics 2008;122:1292–8.
16. Karamlou T, Diggs BS, Person T, et al. National practice patterns for management of adult congenital heart disease. Operation by pediatric heart surgeons decreases in-hospital death. Circulation 2008;118: 2345–52.
17. Menon SC, Cetta F, Dearani JA, et al. Hybrid intraoperative pulmonary artery stent placement for congenital heart disease. Am J Cardiol 2008;102:1737–41.
18. Vida VL, Lago P, Salvatori S, et al. Is there an optimal timing for surgical ligation of patent ductus arteriosus in preterm infants? Ann Thorac Surg 2009;87:1509–16.
19. Lim HG, Bacha EA, Marx GR, et al. Biventricular repair in patients with heterotaxy syndrome. J Thorac Cardiovasc Surg 2009;137:371–9.
20. Hörer J, Schreiber C, Cleuziou J, et al. Improvement in long-term survival after hospital discharge but not in freedom from reoperation after the change from atrial to arterial switch for transposition of the great arteries. J Thorac Cardiovasc Surg 2009;137:347–54.
21. Bové T, De Meulder F, Vandenplas G, et al. Midterm assessment of the reconstructed arteries after the arterial switch operation. Ann Thorac Surg 2008;85:823–30.
22. Petit CJ, Rome JJ, Wernovsky G, et al. Preoperative brain injury in transposition of the great arteries is associated with oxygenation and time to surgery, not balloon atrial septostomy. Circulation 2009;119: 709–16.
23. Sharma R, Talwar S, Marwah A, et al. Anatomic repair for congenitally corrected transposition of the great arteries. J Thorac Cardiovasc Surg 2009;137:404–12.
24. Scherptong RWC, Vliegen HW, Winter MM, et al. Tricuspid valve surgery with a dysfunctional systemic right ventricle. Circulation 2009;119:1467–72.
25. Honjo O, Benson LN, Mewhort HE, et al. Clinical outcomes, program evolution, and pulmonary artery growth in single ventricle palliation using hybrid and Norwood palliative strategies. Ann Thorac Surg 2009;87:1885–93.
26. Tabbutt S, Nord AS, Jarvik GP, et al. Neurodevelopmental outcomes after staged palliation for hypoplastic left heart syndrome. Pediatrics 2008;121;476–83.

27. Grosse-Wortmann L, Yun T-J, Al-Radi O, et al. Borderline hypoplasia of the left ventricle in neonates: insights for decision-making from functional assessment with magnetic resonance imaging. J Thorac Cardiovasc Surg 2008;136:1429–36.
28. Alsoufi B, Al-Halees Z, Manlhiot C, et al. Mechanical valves versus the Ross procedure for aortic valve replacement in children: propensity-adjusted comparison of long-term outcomes. J Thorac Cardiovasc Surg 2009;137:362–70.
29. Cameron DE, Alejo DE, Patel ND, et al. Aortic root replacement in 372 Marfan patients: evolution of operative repair over 30 years. Ann Thorac Surg 2009;87:1344–50.
30. Aalberts JJJ, van den Berg MP, Bergman JEH, et al. The many faces of aggressive aortic pathology: Loeys-Dietz syndrome. Neth Heart J 2008;16:299–304.
31. Yoshimura N, Matsuhisa H, Otaka S, et al. Surgical management of multiple ventricular septal defects: the role of the felt sandwich technique. J Thorac Cardiovasc Surg 2009;137:924–8.
32. Chessa M, Butera G, Negura D, et al. Transcatheter closure of congenital ventricular septal defects in adult: mid-term results and complications. Int J Cardiol 2009;133:70–3.

Key Words: congenital ■ pediatric ■ disease.