There has been a proliferation of publications related to congenital heart disease in major cardiology journals with increased numbers of excellent papers related to this topic. This has made it more difficult to select only a relatively small number of the articles that can be critical to advancements in the field.

As in the past, I have chosen to focus almost entirely on the outcomes of management of the complex conditions that present to surgeons and cardiologists in this field. Sections chosen to report on include patients with tetralogy of Fallot (T/F), transposition of the great arteries (TGA), hypoplastic left heart syndrome (HLHS), Fontan surgery, cardiac surgery in general, adult congenital heart disease (ACHD), and miscellaneous topics. Hopefully, these selections will be useful to those in the field and lead to further research and development of new management strategies that will improve long-term outcomes.

Tetralogy of Fallot

Adamson et al. (1) present a detailed review of all papers related to pulmonary valve replacement (PVR), in T/F, and the question of whether it improves right ventricular (RV) function. These authors reviewed 19 of 730 papers; their search represented the best evidence to answer the question. The data indicate that PVR does improve RV function with associated symptomatic benefit. The authors report consistent reductions in RV end-diastolic and end-systolic volumes with improvement in RV stroke volume. One study also showed improvement in left ventricular (LV) stroke volume. They concluded that PVR improved clinical status and can be performed with low mortality. Unfortunately, there remains a need for a more durable pulmonary valve.

Boni et al. (2) present current strategies in pulmonary valve sparing T/F repair; with evaluation of RV/LV pressure ratios in the European Journal of Cardiothoracic Surgery. Preserving pulmonary valve integrity may offer a better long-term prognosis, despite a risk of residual stenosis. These authors studied 24 patients with T/F who underwent reparative surgery with a valve sparing procedure (median age 8.1 months, range 1.1 to 86.6 months). Intraoperative post-repair echocardiography showed a right ventricular pressure/left ventricular pressure (RVP/LVP) ratio ≥0.70 in 8 patients (33%, group A) and <0.70 in 16 patients (67%, group B). There was neither early mortality, nor functional or rhythm disturbances. One patient required reoperation for residual stenosis at the annular level at 1 year. After a median follow-up of 32.8 months (range 0.6 to 73.1 months), the RVP/LVP ratio decreased by 16% (p = 0.001) in all patients. In group A, the reduction was 28%, and in group B was 12%. A valve sparing strategy reduces pulmonary regurgitation (PR), preserves RV function, and may decrease the incidence of late arrhythmias, which are determinants of long-term outcome. These authors did accept a fairly high RVP/LVP pressure ratio in the operating room. Whether or not this will result in an improved long-term result is still unclear. The operation can be performed with a low mortality as shown here; follow-up studies should help to solve the question of when this operation should be used. Elevated RVP over the long term may result in decreased RV systolic ± diastolic function.

Kanter et al. (3) report on symptomatic neonatal T/F: repair or shunt? These authors studied 37 consecutive, nonrandomized symptomatic neonates with T/F or its variants including complete atrioventricular (AV) septal defect or absent pulmonary valve (but excluding pulmonary atresia) who had either a shunt (n = 17) or primary repair (n = 20). The shunted patients more commonly required emergency operation (24% vs. 0%; p = 0.036); otherwise, the 2 groups were similar. One infant with pre-operative bowel ischemia died 1 day post-operatively after emergency shunting. There were 2 late deaths 11 and 12 months post-operatively, both in primary repair patients. The 16 surviving shunted patients had T/F repair 216 ± 99 days after the original shunt. The shunted patients had shorter intensive care unit and hospital stays for the first operation, which became equivalent when the second hospitalization (repair) values were added. The primary repair patients more frequently had a transannular patch and a tendency to more frequent delayed sternal closure. Four primary repair (20%) and 2 shunted (12.5%) patients required subsequent cardiac operations after complete repair (p = NS). Shunting or primary repair of neonates with symptomatic T/F provided equivalent mortality and results. Shunted patients had fewer transannular patch repairs despite having more emergent initial operations. This suggests that shunt patients...
may do well in terms of getting by with a lower right ventricular outflow tract (RVOT) pressure post-repair and less PR. A selective operative strategy in these infants perhaps can decrease long-term RV dilation and PR.

Transposition of the Great Arteries

Schwerzmann et al. (4) report on ventricular arrhythmias and sudden death (SD) in adults after a Mustard operation for TGA. These authors studied the outcome of 149 adults (mean age 28 ± 7 years) who had undergone a Mustard operation for TGA. During a mean follow-up of 9 ± 6 years, sustained ventricular tachycardia (VT) and/or SD occurred in 9% (13 of 149) of the cohort. Sustained VT/SD was more likely to occur in patients with associated anatomic lesions within New York Heart Association (NYHA) functional class ≥III (hazard ratio [HR]: 9.8, 95% confidence interval [CI]: 3.0 to 31.6) and with an impaired subaortic right ventricular ejection fraction (RVEF) (HR: 2.2, 95% CI: 1.2 to 4.0). There was an inverse correlation of the RVEF and QRS duration. Patients with a QRS duration ≥140 ms were at the highest risk of sustained VT/SD (HR: 13.6, 95% CI: 2.9 to 63.4). Atrial tachyarrhythmia was detected in 66 (44%) patients, but was not a statistically significant predictor of sustained VT/SD. These authors conclude that sustained VT/SD in adults after a Mustard operation for TGA are more common than previously described. Age, systemic ventricular function, and QRS duration are interrelated and are associated with VT/SD. A QRS duration ≥140 ms may help to identify the high-risk patients. Systemic right ventricles do deteriorate with time even after quite successful atrial repair; some of these patients may be candidates for implantable cardioverter-defibrillators.

Rutz et al. (5) report on RV absolute myocardial blood flow (MBF) in complex congenital heart disease. These authors used myocardial contrast echocardiography (MCE) to determine the myocardial microvascular density reflected by relative myocardial blood volume (rBV). The study was conducted to elucidate the relationship between the right ventricular hypertrophy (RVH) and myocardial microvascular changes by quantitative MCE in patients with TGA and T/F. Three groups were included: 22 patients with TGA, 18 patients with T/F, and 22 healthy individuals (controls). MCE was performed at rest and during adenosine-induced hyperaemia. Hyperaemic septal rBV differed significantly between the groups and was the highest in controls and the lowest in TGA. The ratio of hyperaemic to baseline MBF, which differed significantly between the groups, also was lowest in TGA versus T/F and controls. Hyperaemic septal rBV, MBF, and myocardial blood flow reserve (MBFR) showed a significant correlation with RV systolic function as determined by tricuspid annular systolic excursion. These data showed that relative MBF and flow reserve in patients with systemic right ventricles is diminished. This abnormality has been shown previously, but this study provides more data in this regard. Hopefully, there will be research in the future that will provide ways to increase myocardial blood flow, particularly in patients with systemic right ventricles.

Tobler et al. (6) reviewed outcomes in young adult survivors of arterial switch operation (ASO) for TGA. Survival of the 132 infants discharged from the hospital after ASO was 97% and 65 patients (mean age, 21 years) followed at their institution; 17% had at least 1 clinically significant cardiac lesion, including ventricular dysfunction, valvular dysfunction, or arrhythmias. Residual lesions were more common in those who had had cardiac reinterventions in childhood. In adulthood, 5 patients, (8%) had arrhythmia requiring treatment, and 7 patients (11%) required reinterventions (5 reoperations and 2 pacemaker implantations). Intervention for aortic valve regurgitation and aortic root dilatation was not observed. Exercise capacity was reduced in most adults after ASO.

Although most adults after ASO are well, and few have residual defects, there are subgroups who need further cardiac intervention in childhood and who are at higher risk for ventricular ± valvular dysfunction and arrhythmias. The outlook continues to look favorable compared with patients who have had Mustard or Senning repair versus ASO. These patients need follow-up to be sure that their coronary blood flow is adequate and no other residual lesions that will require intervention in the future become evident.

Navabi et al. (7) report on the effect of ventricular septal defect (VSD) enlargement on the outcome of Rastelli or Rastelli-type repair of TGA; 49 patients had Rastelli operations and were involved in a follow-up study divided into 2 groups: group A had VSD enlargement, and group B did not have VSD enlargement. Median age and weight at the time of the operation were 6 years and 17 kg, respectively. The VSD was enlarged in 57% or 28 patients. VSD enlargement was associated with a significantly statistical relation with late ventricular dysfunction, arrhythmia, and residual VSD. Freedom from late death for those without VSD enlargement was 100% at 5 and 10 years of age, 83% at 15 years. Those with VSD enlargement had 74% survival at 5 years and 65% at 10 years. At a median follow-up of 4 years, there were 12 late-onset deaths: 11 patients in the group with VSD enlargement and 1 in the group without. VSD enlargement increased the risk of late death as well as the risk of ventricular dysfunction.

This group represents another way to change the right ventricle to the pulmonary ventricle and the left ventricle to the systemic ventricle with TGA and VSD. These post-Rastelli patients have been shown in the past to have decreased ventricular function and this study shows that VSD enlargement can contribute to this problem.

Hu et al. (8) report on double-root translocation for double-outlet right ventricle with noncommitted ventricular septal defect (DORVncVSD) associated with pulmonary stenosis (PS). These authors report 10 consecutive patients who underwent a double-root translocation procedure at a
mean age of 48 months for DORVncVSD or subpulmonary VSD associated with PS. The VSD was repaired with a patch, and VSD enlargement was done in 3 patients. The aortic translocation was done with (n = 4) or without (n = 6) coronary reimplantation. The neopulmonary artery was reconstructed with a monocusp bovine jugular vein patch (n = 8) or a homograft patch (n = 2). The mean follow-up interval was 22 months. There were no early or late deaths, and no reoperations. Post-operative echocardiography showed satisfactory hemodynamics of the reconstructed biventricular outflow tract and ventricular function. One patient had trivial aortic regurgitation (AR), and 4 patients had trivial or mild PR on follow-up. The early results showed an improved outcome for DORVncVSD or DORV with subpulmonary VSD, associated with PS. Long-term benefits need to be evaluated with a larger number of patients and longer follow-up. This is a long and complex operation; the mean bypass time was 311 min, and cross clamp time was 219 min. Despite these data, there was no significant ventricular dysfunction detected. The standard Rastelli operation, which is the alternative operation, has considerable difficulties with recurrent LV outflow tract obstruction and with ventricular function on follow-up. If this new approach can be accomplished by other groups, it should lead to improved outcomes.

Lim et al. (9) report on biventricular repair for congenitally corrected transposition of the great arteries (CCTGA). These authors studied 167 patients with CCTGA who underwent biventricular repairs. Physiological repairs were performed in 123 patients, and anatomic repairs in 44. Average follow-up was 9 years. Overall survival was 83% at 25 years. In anatomic repair, LV training and RV dysfunction had a negative impact on survival, but bidirectional cavopulmonary shunt had a positive impact. The reoperation-free survival was 10% at 22 years after physiological repair, and 46% at 15 years after anatomic repair. Freedom from any arrhythmia was 49% at 22 years after physiological repair, and 61% at 18 years after anatomic repair. Freedom from systemic AV valve and ventricular dysfunction as well as tricuspid valve and RV dysfunction was significantly higher in anatomic repair than in physiological repair.

Although long-term results of biventricular repair were satisfactory, patients who present with RV dysfunction can be a high-risk group for anatomic repair and selection criteria are particularly important. Anatomic repair is the procedure of choice for these patients if both ventricles are adequate and ventricular dysfunction is only mild to moderate and/or if the surgical technique is modified with the help of an additional bidirectional cavopulmonary shunt. This paper adds to the body of information regarding anatomic repair. This is a long and difficult operation. Retraining of the LV is complex and fraught with danger of biventricular failure. Tricuspid regurgitation (TR) should be treated early, as late replacement of the tricuspid valve is often shown to lead to a marked decrease in RV function. Kollars et al. (10) report on CCTGA associated with TR. This study was performed to determine the effect of LV pressure on RV and LV geometry and the degree of TR. Serial echocardiograms were reviewed from 30 patients with CCTGA who underwent pulmonary artery banding to train the morphologic left ventricle (n = 14, age 0 to 12 years, average 1.1) or left ventricle-to-pulmonary artery conduit placement and ventricular septal defect closure in conjunction with physiological repair (n = 16, age 0.1 to 23 years, median 3.3 years). After pulmonary artery banding, an increase in LV systolic pressure equal to or greater than two-thirds systemic resulted in a decrease in TR from severe to moderate. The percentage of patients with severe TR decreased from 64% to 18%. The RV sphericity index decreased, and the LV sphericity indexed increased. After left ventricle-to-pulmonary artery conduit placement, a decrease in LV pressure equal to or less than one-half systemic resulted in an increase in TR from none to mild. This information was somewhat speculative in the past about the change in TR with the change in LV pressure and septal orientation. This is another way to improve TR in young patients with low LV pressures who can tolerate the pulmonary artery banding. These patients may improve considerably and may not require anything further for many years after this operation if it is successful in decreasing the degree of TR and improving systolic function of the RV.

**Hypoplastic Left Heart Syndrome**

Ohyne et al. (11) present comparisons of shunt types in the Norwood procedure for single-ventricle lesions with systemic ventricular outflow tract obstruction. This is a long-awaited study of patients who were randomly assigned to either the modified Blalock-Taussig (MBT) shunt or the right ventricle–pulmonary artery (RVPA) connection at 15 North American centers. The primary outcome was death or cardiac transplantation 12 months after randomization. Secondary outcomes included unintended cardiovascular interventions, RV size and function at 14 months, and transplantation-free survival until the last subject reached 14 months of age. Transplantation-free survival 12 months after randomization was higher with the RVPA shunt than with the MBT shunt (74% vs. 64%, p = 0.01). The RVPA shunt group had more unintended interventions (p = 0.003) and complications (p = 0.002). RV size and function at the age of 14 months and the rate of nonfatal serious adverse events at the age of 12 months were similar in the 2 groups. Data collected over a mean follow-up period of 33 months showed a nonsignificant difference in transplantation-free survival between the 2 groups.

This is an outstanding achievement to develop this degree of cooperation for a randomized trial in pediatric CV surgery. Continuous surveillance of this cohort is needed to determine whether either of these shunts turns out to be superior over the long term and whether the RVPA group develops ventricular dysfunction with time.
Raja et al. (12) prepared a best-evidence topic in congenital cardiac surgery questioning whether RV-PA shunt for HLHS compared with the MBT shunt is associated with deleterious effects on ventricular performance. General limitations included small numbers, nonrandomized design, and retrospective nature with failure of correlation of echocardiographic indexes to clinical outcomes. The data did show that there was no adverse effect of ventriculotomy on ventricular performance in patients with the Sano shunt in the short and medium terms. It is clear that scars grow with time, and this question remains unanswered in the long term regarding effect on RV performance.

Patel et al. (13) report on the impact of noncardiac congenital and genetic abnormalities on outcomes in HLHS. These authors studied patients diagnosed with HLHS who underwent stage 1 Norwood (n = 1,236), stage 2 superior cavopulmonary anastomosis (n = 702), or stage 3 Fontan (n = 553). In-hospital mortality, post-operative complications, and length of stay were compared at each stage between those with and without noncardiac genetic defects. Stage 1 in-hospital mortality was higher (26% vs. 20%, p = 0.04) with genetic defects. The mean post-operative length of stay was 42 days versus 31 days (p < 0.0001), and post-operative complications were significantly more prevalent in infants with noncardiac genetic defects. Early hazard for death after Norwood was significantly worse in infants with noncardiac defects. Chromosomal defects were highly unfavorable: the early risk of death was doubled, and Turner syndrome accounted for the majority of chromosomal defects in this population. Mode of death was rarely attributable to the noncardiac genetic defect. These data indicate a strong influence of noncardiac abnormalities on survival. Strategies to improve mortality in infants with noncardiac abnormalities should be explored. Presence of chromosomal defects, especially Turner syndrome, should enter decision-management options for patients and physicians.

Vogel et al. (14) reviewed fetal congenital diaphragmatic hernia and LV hypoplasia in terms of outcome; of 125 patients diagnosed prenatally with congenital diaphragmatic hernia (CDH), 111 had a left-sided defect. Of these, 85 were live born, including 20 with congenital heart disease. Gestational age-adjusted dimensions of fetal left heart structures, including aortic valve diameter, mitral valve (MV) diameter, LV long axis, LV short axis, and LV volume were all smaller than normal. On average, the LV contributed 33% of combined ventricular output, lower than the normal, 40% to 50%. The z-scores of left heart structures increased from the pre-natal echocardiogram to the post-natal study, with average changes ranging from 0.56 (aortic valve) to 1.39 (LV volume). Among live-born patients, there was no association between prenatal left heart z-scores and postnatal survival. Hypoplasia of and reduced flow through the left heart are common among fetuses with left-sided CDH. After birth and CDH repair, left heart dimensions generally normalize, with adequate size to support a biventricular circulation, even when there is very low flow through the left heart in mid and late gestation. These data should help physicians to continue to treat these patients aggressively and not to deny their surgery because of their LV size. Outcomes appear to depend more on lung volume, which can correlate with post-natal survival.

McElhinney et al. (15) report on predictors of technical success and postnatal biventricular outcome after in utero aortic valvuloplasty for AS with evolving HLHS; 70 fetuses underwent attempted aortic valvuloplasty for critical AS with evolving HLHS. The median gestational age was 23 weeks. The procedure was technically successful in 52 fetuses (74%). Relative to 21 untreated comparison fetuses, subsequent pre-natal growth of the aortic and mitral valves, but not the left ventricle, was improved after intervention. Nine pregnancies did not reach a viable term or preterm birth. Seventeen patients had a biventricular circulation post-natally, 15 from birth. Larger left heart structures and higher LV pressures at the time of intervention were associated with biventricular outcome. A multivariable threshold scoring system was able to discriminate fetuses with a biventricular outcome with 100% sensitivity and modest positive predictive value. Technically successful aortic valvuloplasty alters left heart valvar growth in fetuses with AS and evolving HLHS and, in a subset of cases, appeared to contribute to a biventricular outcome after birth. Fetal aortic valvuloplasty carries a risk of fetal demise. Fetuses undergoing in utero aortic valvuloplasty with an unfavorable multivariable threshold score at the time of intervention are very unlikely to achieve a biventricular circulation post-natally.

These authors have continued to attempt to modify their selection criteria to determine which patients would be candidates for a biventricular circulation with fetal intervention. The multivariable threshold scoring system takes into account the sizes of the aortic valve, MV, and LV, as well as the LV pressure, and allowed the highly sensitive and moderately specific identification of fetuses that survived post-natally with a biventricular circulation.

**Fontan Surgery**

O’Brien et al. (16) report on the nonfenestrated extracardiac Fontan procedure in a cohort of 145 patients. Hospital survival was 94.5% and takedown of the Fontan was necessary in 4 patients (2%); 3 of these 4 patients were also operative deaths. Although female sex, cardiopulmonary bypass time, and aortic cross-clamp time were found to be significant in the univariate analysis, only cardiopulmonary bypass time and sex were significant in the multivariate analysis for hospital survival. The late survival was 96% (131 of 137 patients) with a mean follow-up of 3.3 years. The freedom from Fontan failure was 92% at 1 year, 89% at 5 years, and 82% at 10 years. Stepwise logistic regression found cardiopulmonary bypass time to be a significant risk factor for late survival. These authors present excellent data...
in terms of going to a standard nonfenestrated extracardiac Fontan procedure. They utilized early extubation in the operating room to enhance cardiac output. Of 8 early Fontan failures, 5 patients expired as a result of inadequate pulmonary capacity. Of the remaining 3, 1 patient died secondary to multisystem organ failure after operative malperfusion, 1 secondary to air embolism, and 1 while awaiting transplantation due to diastolic dysfunction. Further investigations are needed to provide data on pulmonary vascular capacity to further increase the predictive value of preoperative studies. The incidence of supraventricular tachycardia was low at 1 of 128 patients, or 0.8%. Although 16% of the patients lost sinus rhythm in the follow-up period, 38% of that group had an ectopic atrial pacemaker. It is disappointing to see the number of patients requiring home drains for a median time of 21 days. Home treatment of the effusions was apparently well tolerated, and no significant morbidity from this therapy was reported.

Salazar et al. (17) report on “Fenestration During Fontan Palliation: Now the Exception Instead of the Rule.” These authors report 226 consecutive patients who underwent primary Fontan palliation from 2008 to 2009 with the selective use of Fontan fenestration. This technique has evolved with extensive use of an extracardiac conduit with limited cardiopulmonary bypass and decreased fenestration (only 6% of patients had a fenestration used in the last year). Extubation in the operating room was used if at all possible. The mean hospital stay was 10 days. Survival to discharge or 30 days was 98%. Mean age and weight in this cohort was 4 years and 17 kg, respectively. These data support the nonfenestrated Fontan in the majority of patients unless they are high risk. These authors make the decision in the operating room as to whether or not to fenestrate the Fontan patients. Patients known to be high risk such as those with single-lung Fontan palliation, severe AV valve regurgitation, borderline pulmonary vascular resistance or transpulmonary gradient, or with poor ventricular function were more likely to have fenestration. Such high-risk patients were obviously rare, and they may be better served in many cases by transplantation. The discussion after this paper included a comment that operating at high altitude, such as in Colorado, may be a requirement to use fenestration.

Rathod et al. (18) reported that myocardial fibrosis (MF) identified by cardiac magnetic resonance late gadolinium enhancement (LGE) is associated with adverse ventricular mechanics and VT late after Fontan operation. These authors studied 90 patients with a mean age at study of 23 years: 28% had positive LGE, and patients with LGE had lower mean ejection fraction (45% vs. 56%), increased median end-diastolic volume, increased median ventricular mass, higher frequency of regional wall motion abnormalities, and higher frequency of nonsustained ventricular tachycardia (NSVT). In this cohort of late Fontan survivors, MF was common and associated with adverse ventricular mechanics and a higher prevalence of NSVT and SVT. It is clear that patients with definite LGE are a high risk for significant problems with follow-up, including ventricular dysfunction and ventricular arrhythmia. The real question is how to prevent LGE with management of these patients prior to their Fontan operation. Hopefully, early volume unloading and enhanced O₂ saturations will prevent MF in patients being seen now as infants.

Goldstein et al. (19) report on exercise capacity in patients with a single ventricle receiving Fontan palliation. These authors studied 29 Fontan patients and 16 control subjects: Percentage of predicted peak oxygen consumption (VO₂) was lower with supine exercise than with upright exercise, and was decreased in Fontan patients as shown by other studies. Diastolic dysfunction was present in 57% of the Fontan patients and was associated with a reduced percentage of predicted peak VO₂ and supine peak work. Six Fontan patients who underwent supine exercise with indwelling catheters failed to demonstrate the expected decrease in pulmonary vascular resistance characteristically seen with peak exercise. Supine exercise in Fontan patients does not result in an increased VO₂ or oxygen pulse, suggesting that venous return might not be the primary limitation of exercise capacity in this population. Diastolic dysfunction and relatively excessive peak-exercise pulmonary vascular resistance might be more important factors in Fontan exercise limitation. These authors suggest that sildenafil may help patients with Fontan palliation to achieve better exercise performance. It has been suggested that chronic sildenafil use can be advantageous in improving exercise performance in Fontan patients, but previous studies have shown this only with acute administration of the drug.

**Cardiac Surgery (General)**

Huang et al. (20) report on disproportionate growth of the tricuspid valve after decompression of the right ventricle in patients with pulmonary atresia and intact ventricular septum. These authors studied 40 patients who underwent initial RV decompression for planned staged repair. The initial Z-value of the tricuspid valve diameter was obtained the echocardiography-derived normal values. The late Z-value was measured before definitive repair or the last available Z-value, if definitive repair was not yet reached. The mean initial tricuspid Z-value was −6.2 ± 3.5. After treatment, the mean Z-value was −6.0 ± 3.4. Overall, the tricuspid Z-values did not change. Individually, the change in Z-value was larger than +2 in 11 (32%) patients and smaller than −2 in 6. Increases in Z-value were significantly associated with RVP/LVP ratio measured after initial palliation and the initial tricuspid valve Z-value.

These findings support the possibility of neonates with very small tricuspid valves undergoing biventricular repair after RV decompression surgery. There have been previous studies suggesting that tricuspid valves prior to attempting biventricular repair would have to be ≥−5.
These authors found a number of patients who became candidates for biventricular repair despite the initial Z-values of $\leq -6.3$. These data suggest that a very small tricuspid valve may not be an indication for abandoning possible biventricular repair. The question remains as to whether one can make the atrial defect slightly smaller with the initial RVOT opening to “force” blood across a small tricuspid valve and pulmonary outflow to enlarge the RV long term. Obviously, this is a risky procedure that should be quantified in terms of the ability to pursue this strategy in the long-term.

**Surgery for Ebstein’s Anomaly**

Malhotra et al. (21) report on selective RV unloading and novel technical concepts in surgery from Ebstein’s anomaly. These authors report on 57 non-neonatal patients who underwent Ebstein’s anomaly repairs at a median age of 8.1 years. All were symptomatic in NYHA functional class II to IV. Pre-operative, 26 had mild to moderate cyanosis at rest, and a number of valve reconstructive techniques were used that differed substantially from those currently described. Bidirectional Glenn (BDG) was performed in 31 patients (55%) who met specific criteria. No early or late deaths occurred. At the initial repair, 3 patients received a prosthetic valve. Four patients required reoperation for severe TR, and repeat repairs were successful in 2 patients. At follow-up (3 months to 6 years), all patients were acyanotic and in NYHA functional class I. TR was mild or less in 49 (86%) and moderate in 6 (11%). Freedom from prosthesis was 91% (52 of 57). Following a protocol using BDG for ventricular unloading in selected patients, these authors report a durable valve sparing repair using the techniques described in terms of valve reconstructive surgery including the “play it where it lies” approach to the tricuspid valve in which the reconstruction is performed at the functional orifice instead of moving the valve to the anatomic tricuspid annulus. Avoidance of detachment and reimplantation of valve leaflets and a limited plication performed only at the level of the displaced valve rather than complete plication of the entire atrialized RV was used.

The question as to whether RV systolic function is really normal in any patients with Ebstein’s was brought up in the discussion of this paper. Hopefully, these authors will use exercise data on further follow-up of their patients to show evidence of good, long-term results.

Shinkawa et al. (22) report on the management and long-term outcome of 40 symptomatic patients with Ebstein’s anomaly in the neonatal period. There was no early intervention required in 16 of the 40 patients with a hospital survival of 94% (15 of 16) and no late mortality. The remaining 24 patients underwent surgical intervention in the neonatal period; a shunt alone was performed in 9 patients with an actuarial survival of 88.9% at 1 year and 76.2% at 5 and 10 years. For the patients undergoing surgical intervention, survival estimates for the 11 patients with an RV exclusion procedure were 64% at 1, 5, and 10 years, and 48% at 15 years compared with 25% at 1, 5, and 10 years for the 4 patients with tricuspid valve repair. All long-term survivors were in NYHA functional class I or II, and only 1 patient required ant arrhythmic medication. These authors suggest that optimally, neonates with symptomatic Ebstein’s anomaly can be successfully managed with only early prostaglandin without surgical intervention. For those patients who cannot be weaned from prostaglandin, a systemic/pulmonary (SP) shunt alone can be used if heart failure is absent. For patients with both cyanosis and heart failure necessitating tricuspid valve intervention in the newborn period, the data suggest that RV exclusion (tricuspid valve closure with an SP shunt) results in the most favorable outcome. Although tricuspid valve repair allows for biventricular anatomy and remains an important part of the surgeon’s armamentarium for older children, it is best reserved for those neonates with the most favorable anatomy based on pre-operative and intraoperative assessment.

**Aortic Valve/Aortic Disease**

Oliver et al. (23) report on risk of aortic root or ascending aorta complications (AACs) in patients with bicuspid aortic valve (BAV) with and without coarctation of the aorta (COA). These authors report on 631 patients in whom BAV was diagnosed by echocardiography or surgical inspection. These were further subdivided into 2 groups according to presence of an associated COA, AACs including aortic aneurysms (ascending aorta $\geq 55$ mm), and aortic dissection, rupture, or decreased perfusion. Patients with a BAV and COA had a greater prevalence of AACs than those with an isolated BAV. The coexistence of COA was the only significant predictor of AACs (odds ratio: 4.7, 95% CI: 1.5 to 15; $p = 0.01$). The clinical and echocardiographic data were reviewed for 341 patients with BAV without an AAC at baseline who had undergone serial examination $>1$ year apart. The mean follow-up was 7 years (interquartile range was 3.5 to 10.2; total, 2,436 patient-years). A new AAC occurred in 13 patients, and the incidence of AACs was 1.3/100 patient-years in the COA group versus 0.2/100 patient-years in the non-COA group. All acute aortic events at follow-up occurred in patients with BAV and COA. The long-term incidence of AACs in patients with isolated BAV is low, but patients with BAV and associated COA are at increased risk. These authors have shown that a BAV rarely has major AACs unless there is a COA present. Prior studies have shown that BAV and the tricuspid aortic valve are equal in prevalence with surgery in older patients with AACs. There is an ongoing debate about whether all patients with BAV have AACs that are genetic in origin or are due solely to hemodynamic stress imposed on the valve. In this setting, surgical recommendations for patients with Marfan syndrome might not be applicable to patients with BAV. The incidence of acute AACs found in the present...
study gives support to this consideration unless coarctation is present.

**Other Important Papers**

Lin et al. (24) report on a retrospective review on of 922 patients who had repair of VSD, AV septal defect, or T/F with a median follow-up of 4.1 years. The incidence of delayed AV conduction block was 0.3% to 0.7%. Postoperative AV conduction block developed in 21 of the 922, being transient, with return of conduction 3 days (1 to 14) after surgery in 13 and permanent, with pacemakers implanted 10 days after surgery, in 8. There were 905 patients at risk for delayed AV conduction block, 3 had second- or third-degree block at 2, 8, and 16 months after surgery. Two of these 3 had transient post-operative block. For isolated VSDs, the incidence was 1 of 496 (0.2%). There were 8 late deaths at 31 months (7 to 45 months) after surgery. Five had normal conduction at death, but for 3 patients, the conduction status at death could not be determined. Including these 3 patients as possible cases of delayed AV block yields an incidence of 0.3% to 0.7%. The incidence of early AV conduction block requiring a pacemaker was 0.9% and that of delayed AV conduction block was 0.3% to 0.7%. Transient AV conduction block may be a marker for increased risk of delayed block. These data indicate that block after surgery for VSD is much less than recently reported for percutaneous VSD closure.

Shuhaiber et al. (25) report intraoperative assessment after pediatric cardiac surgical repair: initial experience with C-arm angiography. Hybrid operating rooms have been increasingly used in selected high-risk patients. Angiograms in the operating suite can offer useful data in selective studies that echocardiograms cannot duplicate. These authors performed intraoperative angiographic analysis in 18 cases. Cases in which intraoperative surgical management can be influenced by the results of the on-table angiograms include complex shunt patency, coronary patency, pulmonary artery anatomy, and conduit compression. Angiograms can be useful in certain complex patients and may well become a standard in the future in these types of patients to avoid the time to confirm or refute any outstanding concerns during surgical intervention. Intraoperative angiography should reduce the need for early post-operative catheterizations by alerting the surgeon to potential or actual surgical concerns in a timely fashion.

**Adult Congenital Heart Disease**

Opotowsky et al. (26) report on trends in hospitalizations for adults with ACHD in the U.S. These authors used nationally representative data from 1998 to 2005 to identify patients ≥18 years of age admitted to an acute care hospital with an International Classification of Diseases-9th Revision code designating a CHD diagnosis. National estimates of hospitalizations and total hospital charges by year were calculated. The number of ACHD hospitalizations increased from 35,992 ± 2,645 in 1998 to 72,656 ± 5,258 in 2005. During this period, the annual number of admissions grew for both simple and complex diagnoses. The percentage of admissions originating in the emergency department (42%) or involving cardiac surgery (18%) remained stable during the study period. The average patient age and proportion of patients with ≥2 medical comorbidities also increased. Mean hospital charges per hospitalization increased by 127%, and the estimated total national charges for these hospitalizations increased by 357% from $691 million in 1998 to $3.16 billion in 2005. The number of hospital admissions for ACHD in the U.S. more than doubled between 1998 and 2005. Hospital charges attributed to these admissions have grown even more dramatically. This is a primary result of several unrelated phenomena, most notably improvements in pediatric care and noninvasive diagnostic testing. As a result of the increased number of adult patients with simple and complex CHD, health care resources are straining to meet the needs of further planning for the care of these patients both in terms of facilities and physicians is urgently needed.

Verheugt et al. (27) reported on all hospital admissions from 2001 to 2006 of 5,798 adult patients with congenital heart disease with the mean age of 39 years from the Dutch national registry. Admission rate in these patients was high and exceeded the general Dutch population by 2 to 3 times; this difference was more pronounced in the older age groups. Altogether there were 8,916 admissions, 5,411 of which were for cardiovascular indications. Among cardiovascular admissions, referrals for arrhythmias were most common at 31%. Of 4,926 interventions, 2,459 were cardiovascular, most often reparative interventions or cardioversion. Most noncardiovascular admissions were obstetric. Among defects, univentricular heart and tricuspid atresia had the highest incidence and duration of admission. These authors again show the health care utilization and registry in the adult patients with congenital heart is high and increases with age. Admission rates are at least 2 times higher than in the general population, and most marked in the older age groups.

Verheugt et al. (28) report on the mortality in ACHD from the Dutch registry. Of 6,933 patients, 197 died during a follow-up of 24,865 patient-years as compared with the general population, there was excess mortality, particularly in the young. Median age of death was 48 years. Of all deaths, 77% had a cardiovascular origin; 45% were due to chronic heart failure or sudden death. Age predicted mortality, as did gender, severity of the defect, number of interventions, and number of complications. Several complications predicted all-cause mortality beyond the effects of age, sex, and congenital heart disease severity (i.e., endocarditis, supraventricular arrhythmias, ventricular arrhythmias, conduction disturbances, myocardial infarction, and pulmonary hypertension). These risks were similar in patients above and below 40 years of age. Almost all complications predicted death due to heart failure; conduction distur-
bances and pulmonary hypertension predicted sudden death. The vast majority die from cardiovascular causes. Complications are equally hazardous in younger as in older patients. This is another wake-up call in terms of these patients. These patients need physicians who are acquainted with their anatomy and physiology and can provide the care needed. There is a need for training of these physicians in all corners of the world.

Miscellaneous

Kim et al. (29) report on successful repair of atrial septal defect (ASD) with Eisenmenger syndrome after prolonged sildenafil therapy. This patient underwent repair of an ASD, after treatment for 2 years with sildenafil and has been monitored for 4 years after repair. This case supports a "treat and repair" approach using advanced pulmonary vasodilator therapy that may be optimal in selected patients with otherwise inoperable severe pulmonary hypertension. This case can raise hope to some of these patients with severe symptoms secondary to pulmonary arteriopathy and associated CHD.

Ionescu-Ittu et al. (30) report on prevalence of severe congenital heart disease after folic acid fortification of grain products, with a time trend analysis in Quebec, Canada. These authors report on data analyzed in 2 time periods, before and after fortification of grain products, specifically flour and pasta products with folate, in which they studied the prevalence of severe congenital heart defects including T/F, endocardial cushion defects, univentricular hearts, truncus arteriosus, or transposition complexes in the Quebec administrative databases. Among the 1,324,400 births in Quebec in 1990 to 2005, there were 2,083 infants born with severe congenital heart defects, corresponding to an average birth prevalence of 1.57/1,000 births. Time trend analysis showed no change in the birth prevalence of severe birth defects in the 9 years before fortification (rate ratio: 1.01, 95% CI: 0.99 to 1.03), whereas in the 7 years after fortification, there was a significant 6% decrease per year (0.94 to 0.97). Public health measures to increase folic acid intake were followed by a decrease in the birth prevalence of severe congenital heart defects. These findings support the hypothesis that folic acid has a preventive effect on heart defects. Hopefully, other data can be found to substantiate this initial observation.

This is the first study, to my knowledge, to show that changes in intake of this common vitamin can change the incidence of congenital heart disease. It certainly has worked for neural tube defects and, hopefully, can be shown in similar studies to have the same effect in congenital heart disease.

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