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The unique challenge of treating the adult patient with congenital heart disease

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For the most part, adult patients with congenital heart (ACH) disease are "products" of pediatric medical and surgical advances that took place in the second half of this century. Until recently, these patients received their care *from cradle till coffin* from pediatric caregivers because it was felt that they faced an adult world not yet fully prepared for their care. Growing interest and need over the past few decades has led to the development of a number of ACH programs that centralize care in a handful of medical centers of expertise. These centers offer coordinated medical, surgical, catheter-based, and specialty care for ACH patients and their families. Throughout their adolescent and adult years, ACH "survivors" present to their caregivers with medical and surgical problems that are different from the ones they experienced in their pediatric years. Novel approaches to medical, surgical, and catheter-based diagnosis and therapy have led to improved well-being and prolonged survival. As "untreated" and "treated" natural history patterns emerge, caution is advised to avoid extrapolations and assumptions based solely on congenital pediatric or acquired adult heart disease principles. In this presentation, I will review the indications, therapeutic interventions, and results of catheterization-based management of particular illnesses in ACH. Extension of these interventions and techniques to adult patients with acquired noncongenital heart disease will also be discussed.

The ACH patient

Every patient with congenital heart disease is unique from an anatomical, physiological, medical, and surgical point of view. An in-depth understanding of each of these aspects is requisite before contemplating potential etiologies of decompensation and approaches to therapy. As well, an extensive review of a patient's electrocardiography, radiography, echocardiography, and nuclear testing (lung perfusion scan) results is mandatory prior to formulating management plans.

The majority of presenting complaints in ACH patients are the sequelae of ventricular dysfunction, atrial or ventricular arrhythmias, or pulmonary vascular disease. These patients face increasing and cumulative hemodynamic effects from surgical scarring and insufficient myocardial preservation during prior surgery, uncorrected abnormalities in ventricular pre-load and after-load, alteration in red blood cell mass and tissue perfusion, and the progressive decrease in myocardial compliance with aging. These effects contribute to increasing systolic and diastolic ventricular dysfunction and arrhythmia, and also contribute to elevation in ventricular filling pressures and volume, with atrial enlargement, sluggish atrial flow and thrombosis, and development of atrial arrhythmia. Additionally, in some patients, inadequately controlled pulmonary blood flow may lead to increasing incidence of pulmonary vascular disease, *in situ* thrombosis, and after-load effects on pulmonary ventricular function. These physiologic issues must be placed within the context of particular anatomic lesions as outlined below.

Secundum-type atrial septal defects

After bicuspid aortic valve disease, secundum-type atrial septal defects (ASD-2) are the most common cause of congenital heart disease in adults. Patients with unoperated or previously unrecognized ASDs are therefore not infrequent visitors to the cardiologist's office. As physicians, we rely on echocardiography for physiologic confirmation of chronic excessive right ventricular volume loading rather than catheterization-based documentation of a "hemodynamically significant" shunt (pulmonary/systemic flow ≥1.5) at a single moment (Figure 1). We therefore do not recommend "routine" catheterization of these patients for determination of intracradiac shunting. Excessive shunt correlates with risk for development of dyspnea, congestive failure, atrial arrhythmias, and less commonly, pulmonary hypertension or paradoxical embolization.

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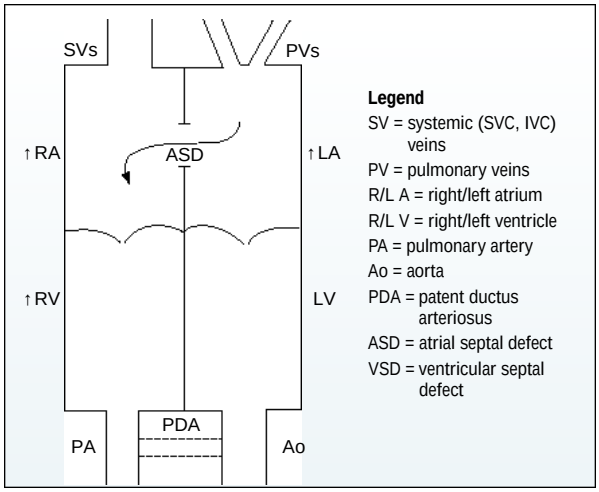
Figure 1: ASD physiology. Intracardiac shunting is governed by relative resistance to ventricular filling. Unless RV function is compromised, flow is left to right with enlargement of right-sided chambers.



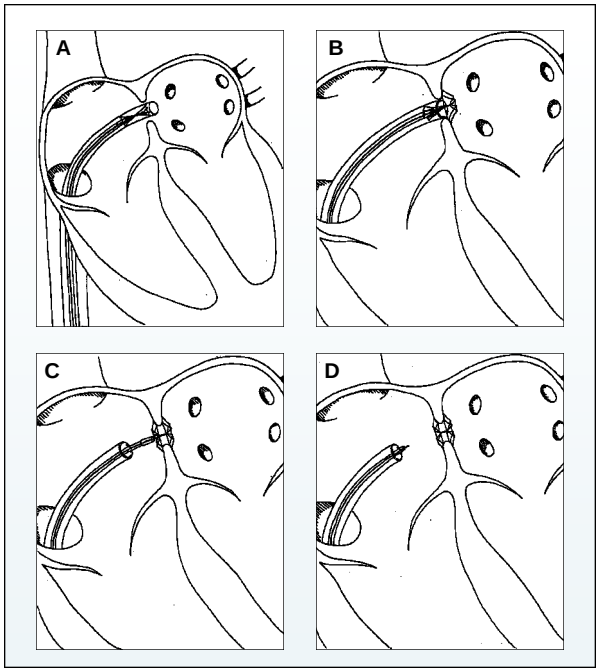
Surgical patch closure or primary suture of ASD-2 via full or "mini" sternotomy or thoracotomy remains one of the safest, most effective, and most cosmetically acceptable adult cardiac surgical procedures.[1,2] However, significant perioperative morbidity may still ensue. Increasing age (with its attendant medical co-morbidities) and pulmonary hypertension are independent risk factors for increased surgical mortality. Minor "patch-margin" shunting, detectable by Doppler echocardiography, is reported to persist in 7% to 8% of patients who are treated surgically. The incidence of major or minor neuropsychiatric complications after cardiopulmonary bypass in this population has refocused attention on the potential for transcatheter closure techniques.

The atrial septum can be readily crossed from femoral venous access, and the placement of a large-caliber guiding sheath allows extrusion of compacted expandable devices in the exact location (Figure 2). The majority of septal defect closure devices are designed as locked or attachable opposing structures that engage the rim of the tissue surrounding a central hole in the heart wall.[3-8] Recently, "self-centering" devices, based on filling the defect with material flared on ether side to buttress the device, have been developed.

Autopsy series from the past confirm that the median atrial septal defect size in adults is approximately 2 cm. Early devices such as the Bard clamshell septal occluder relied on sufficient device size (approximately 1.8 to 2.2 times the maximal balloon catheter stretched defect size) to allow for adequate gripping and seating of the occluder. A maximal device size of 40 mm allowed for closure of the majority of centrally located secundum-type ASDs. Newer, "self-centering devices"may be able to close substantially larger defects.

Clinical experience has been greatest with the Bard clamshell septal occluder and the Sideris standard buttoned device. The technique of deployment in adults is similar to that in the pediatric patient. Of the initial 35 adult patients (aged 18-76 years) undergoing transcatheter ASD-2 dosure with a Bard clamshell septal occluder in Boston, 21 (60%) had a significantly increased risk of operative morbidity or mortality. All 32 patients with appropriately sized defects (≤27 mm maximal stretched diameter) had stable device implantation on the atrial septum. Five patients had significant leaks (device arm herniation across the atrial septum at time of deployment in three

Figure 2: Technique of transcatheter deployment of a Clamshell occluder for secundum-type atrial septal defect closure.



and implantation of an inappropriately small device in two). These defects were subsequently closed by implantation of a second device (3) or via surgery (2). No embolic events or bouts of bacterial endocarditis occurred in 42 to 67 months of follow-up. Minimal residual shunting detected by Doppler echocardiography was present in 32% of patients at one year of follow-up. The significance of this degree of residual shunting is unknown and requires further study, but appears benign at this time.

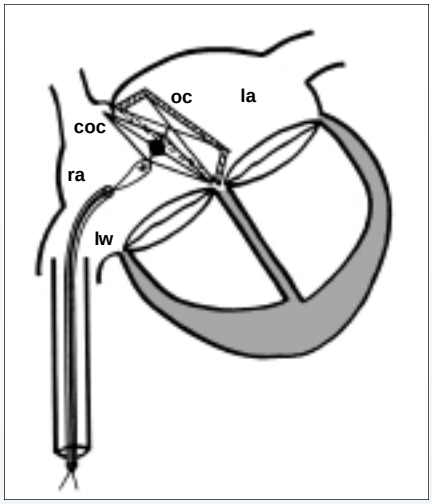
Occult device arm fractures, typically at the spring-loaded hinge point of the device arms,have been seen with larger incidence with increasing device sizes (>50% incidence with devices ≥28 mm in diameter). These fractures have contributed to residual shunting or the development of granulation tissue in areas of contact between the device arm and the atrial wall in <1% of patients with implanted devices. A lower incidence of arm fractures is expected with the CardioSeal modification, which is undergoing clinical trials for low-risk (with <20 mm stretched diameter) and high-risk patients.

Immediate and intermediate-term ASD-2 closure success has been reported with use of the Sideris buttoned device (Figure 3) which has a 12-month actuarial event-free survival of 89% in all pediatric and adult patients. Wire abnormalities, device unbuttoning, arm fractures, and atrial perforations have been reported and are felt to be more common with larger sized defects and devices.[6] Longer-term follow-up is awaited. In limited series, the centering buttoned device, the ASDOS system, and the Amplatz self-centering device have been used successfully to close defects of up to 35 mm in maximal diameter; however, trials with these devices are ongoing. More advances in prosthesis design may eliminate current limitations of transcatheter closure of routine ASD-2.

Recommendations

Based on the presence of right ventricular volume overload and lack of concomitant confounding intracardiac

Figure 3: The "buttoned" double-disc prosthesis.



Legend:
oc = occluder disc
coc = counteroccluder disc
la = left atrium
ra = right atrium

pathology, we recommend closure of atrial septal defects. Moderate-sized secundum-type defects are amenable to transcatheter device closure. At the present time, however, all such devices remain investigational. Procedural risks, in large part, remain a factor of operator experience and appear minimal in centers of expertise. Longer-term risks appear acceptable, making transcatheter ASD closure of moderate sized (<2 cm maximal balloon catheter stretched) ASD-2 a reasonable alternative to current surgical closure. While all available devices appear to have similar efficacy for moderate-sized holes, experience remains greatest with the clamshell devices (CardioSeal modification).

Patent foramen ovale (presumed paradoxical embolism)

Therapeutic closure of patent foramen ovale (PFO) in patients with stroke and presumed paradoxical embolism (PPE) remains controversial, given that the cause and natural history of these embolic events are unclear.[10-12] Recent evidence in 140 consecutive patients suggests an infarct or transient ischemic recurrence rate of approximately 2% and 4%, respectively, regardless of antiplatelet or anticoagulant therapy.[10-12] The significance of whether clinical events (multiple prior embolic or silent events by brain scanning or events that occur with Valsalva straining) or echocardiographic events ("large volume" right-to-left shunting,atrial septal aneurysm) are high risk characteristics for recurrence of PPE is unknown. However, a single, recent, risk-benefit analysis recommends PFO closure,either surgically or via a transcatheter approach, in such patients.[13] Published surgical experience suggests the potential for PFO closure since there was no recurrence of PPE in 32 patients at mean follow-up of 19 months. This literature is counterbalanced by the potential for significant morbid outcomes as reported in a recent series of similarly treated patients.

Our experience with Bard clamshell septal occluder transcatheter PFO ± atrial septal aneurysm closure in 38 patients with PPE revealed successful implantation in all patients, minimal residual shunting (as detected by Doppler echocardiography) in 21%, and no recurrent cerebral infarcts at mean follow-up of 37 months.[14] Two patients (one with residual shunting, one with left atrial granulation tissue near a broken device arm) had transient neurologic ischemia. Both underwent surgical device removal and PFO closure without complication. Published experience with PFO closure for PPE utilizing the buttoned device is similar, with no recurrence of PPE in 5 patients at mean follow-up of 30 months.[15]

Recommendations

Experience with PFO closure is greatest with clamshell-type devices (including the CardioSeal modification) and the buttoned Sideris device, both of which appear best suited anatomically for successful closure of the foramen. At present, given the lack of convincing long-term natural history data regarding embolic recurrence (with and without medical therapies), as well as limited follow-up data after transcatheter PFO device closure with differing controls in the two groups, we currently recommend either correction of an underlying procoagulation abnormality or six months of oral anticoagulation (target INR >2.5). This advice is based on recurrence data for venous thrombosis and embolization, with subsequent recommendation based on relative patient risk. There is an ongoing trial comparing oral anticoagulation with deployment of a CardioSeal device during the initial period after "first event."We recommend device implantation for patients who enroll in similar protocols to determine optimal therapeutic strategies. "Compassionate" device implantation should be reserved for patients with an absolute contraindication to medical therapy or with multiple recurrences of embolic phenomenon.

Patent foramen ovale and hypoxemia

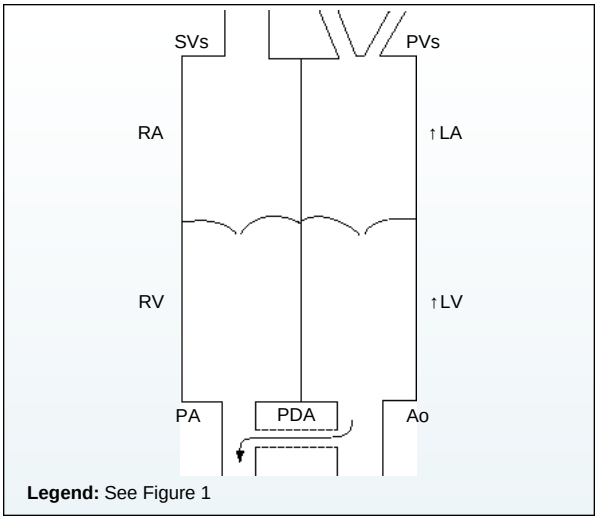
Pressure overload of the right atrium may lead to pathologic right-to-left shunting in patients with a PFO. This may accompany chronic alteration of right-sided filling or capacitance (right ventricular infarction, pulmonary embolism, Ebstein anomaly), or transient alteration in right ventricular filling seen with changing from a supine to an upright position (orthodeoxia-platypnea syndrome). We have closed PFOs with a Bard clamshell septal occluder or CardioSeal device in more than 30 adults with right ventricular infarctions, Ebstein anomaly, and the orthodeoxia-platypnea syndrome.[16] In all but two patients with Ebstein anomaly, catheter closure relieved cyanosis and symptoms, and eliminated the need for open-heart surgery. The long-term benefit of PFO closure in patients with Ebstein anomaly requires further study.

Patent ductus arteriosus

Although under debate in the modern era, an audible patient ductus arteriosus (PDA) in adults is felt to be a marker of increasing risk for development of bacterial endocarditis,and if accompanied by moderate volume shunting, a marker for left ventricular dysfunction (Figure 4). Surgical PDA closure in adults may be more complicated than in children because of anatomic features such as calcification, increased friability, aneurysmal dilation (use of intravascular ultrasonography may assist in identification), and increased risk of multiple organ system comorbidity. Trancatheter techniques have been attempted to offset adult surgical risk,[17-21] utilizing the:

- Ivalon plug (Europe and Japan)
- a similar botallo occluder (Moscow)
- Rashkind and Bard double umbrella occluders
- buttoned device
- embolization coils (routine, screw-apart, bagged)

Figure 4: PDA physiology. Intravascular shunting is governed by relative resistance of pulmonary vs. systemic arterial bed. Unless pulmonary vascular disease exists, flow is left to right with enlargement of left-sided chambers.



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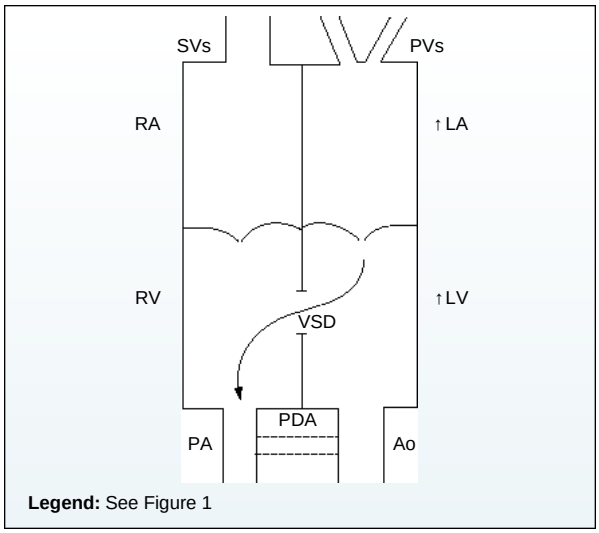
Our initial experience with Rashkind or USCI patent ductus occluders for attempted PDA closure in 21 adults revealed safe implantation, regardless of shape, calcification, presence of pulmonary hypertension, or congestive heart failure. Similar findings in larger numbers of adults with PDA (USCI patent ductus occluder, n=55; Rashkind occluder, n=51) have confirmed these results for PDA of all shapes from 2 mm to 13 mm in size. Incidence of a 50% echocardographically-detectable residual shunt can be detected immediately after device implantation, decreasing to approximately 20% to 25% six months after device implantation. Nearly 100% occlusion can be accomplished in all patients with implantation of additional devices or embolization coils.

The bulky nature and cost of umbrella closure devices for PDA has led to growing experience with transcatheter occlusion of PDAs of all sizes with embolization coils.[22-26] Our initial hesitation in using this technique for larger-sized PDAs (4-7 mm minimal diameter) has been offset by recent advances in coil deployment strategies, including simultaneous placement of multiple coils,use of detachable "screw-apart" retractable or bagged coils to ensure appropriate positioning, prevention of coil embolization during deployment and by entrapment with a balloon catheter within the aortic ampullae during the initial phase of deployment, and by combined bioptome-snare manipulation from either end of the coil.[27-29]

At the Boston Adult Congenital Heart (BACH) Service, we have successfully occluded PDAs ≤7 mm minimal diameter with ≤7 embolization coils in patients ≤60 years. Introduction of longer embolization coils (0.050 inch wire diameter) may allow for successful closure of larger PDAs with fewer coils. The variability in size and shape of PDAs will likely result in the occasional need for umbrella device implantation and appropriate expertise should be maintained. At BACH, we close all audible PDAs in adults via a transcatheter approach; patients should not leave the catheterization laboratory with evidence of residual ductal flow. Bacterial endarteritis precautions are maintained until no residual echocardiographic shunt is detectable. Repeat transcatheter PDA closure should be performed as needed. Recent cost comparisons between transcatheter and surgical PDA occlusion in pediatric patients do not address issues of surgical risk and co-morbidities in the ACH patient and are, therefore, nonapplicable.

Figure 5: VSD physiology. Intracardiac shunting is governed by relative resistance to ventricular contraction and less so to ventricular filling. Unless RV function is significantly compromised or severe pulmonary hypertension/vascular disease exists, flow is left to right with enlargement of left-sided chambers.



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Recommendations

Despite unclear risks of bacterial endocarditis in the current antimicrobial era, we recommend transcatheter PDA occlusion as the procedure of choice for adults with audible PDA and PDA associated with left ventricular dysfunction.[30,31] Coil embolization is successful for nearly all patients, with occasional use of alternative occlusion devices for the uncommon extremely large PDA without associated pulmonary vascular disease.

Ventricular septal defect

Transcatheter closure techniques have been applied to the treatment of congenital and acquired ventricular septal defects (VSDs) in adults (Figure 5) in an attempt to eliminate the need for, or reduce the risk and complexity of, surgical repair.[32,35] In what is perhaps the most technically demanding of interventional catheter procedures performed in our laboratory, we have successfully offered closure to patients with acquired ventricular septal rupture (VSR) after myocardial infarction (MI) or with congenital or postoperative residual muscular VSDs anatomically distant from the aortic valve.

While recent surgical advances have dramatically improved the short- and intermediate-term survival of adults with VSR after MI, operative risk remains substantial and may be compounded by the location of the VSR, the presence of right or left ventricular dysfunction, multiple organ system failure, medical co-morbidities, or prior incomplete surgical attempt at repair.

Since February 1990, we have utilized either the Bard septal clamshell occluder or the CardioSeal device in attempts to limit VSR after MI in:

- 7 patients without prior surgical repair who were felt to have prohibitive surgical risk. Survival past hospitalization or to the present occurred in 3, all of whom had presented months after initial VSR. All survivors are in NYHA class II.
- 11 similarly ill patients who had postoperative residual patch-margin defects after attempted VSR closure. Survival

(mean 54 months) in these patients was not limited by procedural success.

In these patients, septal rupture was often associated with a wide (18-21 mm) necrotic "lake" within the septum, while defects in the patch margin ranged from 8-25 mm by maximal balloon stretching. To date, however,we have not encountered a defect that was not anatomically or procedurally amenable to device implantation. We expect future trials of a more aggressive combined transcatheter-surgical strategy with intense medical-surgical collaboration in an attempt to offer prolonged improvement in those patients felt to be at extreme surgical risk.

Recommendations

At present, closure of a VSR after MI appears most successful when orchestrated by a combined surgical-medical team employing a strategy of expeditious primary surgical repair followed by transcatheter device closure of a residual defect, as required. Because of the technical demands, transcatheter device closure remains limited to few centers such as the BACH. Given current device limitations, a strategy of primary transcatheter VSR repair, regardless of surgical risk or co-morbidities, remains unfounded.

Postoperative residual defects, collaterals, and fenestrations

Embolization coils and umbrella-type devices have been utilized in adults (as in children) at high or prohibitive surgical risk to successfully eliminate:

- residual central aorta or systemic artery to pulmonary artery shunts or collaterals
- systemic venous to pulmonary artery or pulmonary venous shunts or collaterals
- interatrial baffle communications or iatrogenic Fontan fenestrations
- left superior or inferior vena caval connections to the left atrium
- coronary artery fistulae
- systemic and pulmonary arterio-venous malformations
- paravalvar leaks

The role of combined surgically fenestrated repair of intracardiac defects (ASD, VSD, PDA) associated with pulmonary vascular disease in concert with subsequent long-term pulmonary vasodilator therapy and ultimate transcatheter fenestration closure remains to be defined.

Valvar aortic stenosis

The acceptance of balloon aortic valvuloplasty (BAV) as palliation for children with congenital valvar aortic stenosis (VAS) is in contradistinction to the treatment of elderly patients with calcific VAS.[36] Two recent reports have underscored the utility of BAV in selected young and intermediate-aged adults with noncalcified stenotic aortic valves.[37,38]

At BACH, 18 adults (aged 17-40 years) with congenital VAS (15 with identified valvular dysmorpholgy, 2 bicuspid valves, 3 unicuspid valves) underwent BAV with balloons chosen to be 90-100% the diameter of the aortic annulus.[37] Immediate procedural success was achieved in 16/18 patients and yielded a decrease in peak systolic ejection gradient from 85 to 38 mm Hg, without periprocedural death, MI, or recognized embolization. Only 1/11 had an increase in degree of aortic insufficiency after BAV classified as ≥mild-moderate. During a mean follow-up of 38 months, 5 patients required aortic valve surgery. Patients with increased valvular calcification demonstrated a trend toward higher gradients both before and after BAV, and decreased incident-free survival compared to patients without calcified valves.

Sandhu and colleagues reported similar short- and intermediate-term transvalvar gradient reduction with BAV (from 73 mmHg before BAV to 35 mm Hg immediately after dilation) in 15 younger (16 to 24 years) patients with congenital VAS (12 bicuspid).[38] Only 2/6 had increase in degree of aortic insufficiency after BAV classified as ≥ mild-moderate. During mean follow-up of 18 months, 3 patients required aortic valve surgery. Correlation of BAV success to degree of valvar calcification in this younger aged population was not examined.

Recommendations

The above findings support BAV for noncalcified congenital VAS (predominantly bicuspid disease) in young- and intermediate-aged adults. This procedure can provide effective palliation and prolong the interval to surgical intervention without significantly increasing cardiac morbidity or serious complications. Risk and success of surgical repair for the uncommon patient sustaining avulsion of a valvular cusp during BAV does not appear compromised by attempted BAV if echocardiographic evaluation and surgical therapy are accomplished in timely fashion. The impact of surgical pulmonary autograft on the role of BAV for congenital VAS has not been evaluated. We currently recommend an attempt at valvuloplasty for symptomatic patients age ≤40 years with a bicuspid valve associated with gradients ≥60 mm Hg.

Valvar pulmonary stenosis

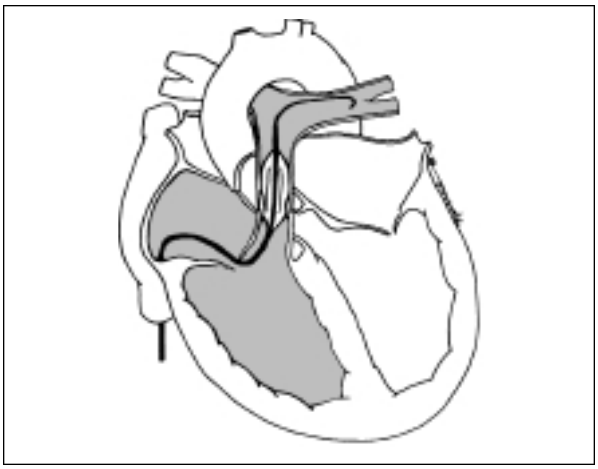
The First and Second Natural History of Congenital Heart Defects Studies reviewed the course and treatment of valvar pulmonary stenosis (VPS) primarily in infants and children,[40] however, the natural history of VPS in older patients is less well-defined. Balloon pulmonary valvuloplasty (BPV) has become the treatment of choice for young patients with VPS and results in immediate success in 80% to 98% of 784 procedures. Transvalvar gradient reduction ranged from 85 mm Hg to 33 mm Hg, with 8% of patients requiring repeat BPV, surgical valvectomy, valvotomy, or right ventricular outflow enlargement during mean follow-up of 33 months in pooled or registry data (Figure 6).[41,42] Long-term success of BPV has been shown to correlate with valve morphology, valve annulus size, balloon-to-annulus dimension ratio, and immediate hemodynamic results. These combined results contain data from only 35 patients over the age of 20 years.

Numerous recent single-center reports of 4-53 patients have demonstrated similar immediate gradient reduction with BPV for VPS in young and middle-aged adults, (13-55 years), utilizing standard single, Inoue, or double balloon techniques to achieve a balloon-annulus dimension ratio between 1.1 and 1.4.[43-47] Our experience at BACH in over 30 adults (18-72 years), with VPS of nondysplastic valves treated with BPV, with a mean follow-up of 8 years, confirms long-term diminution in transvalvar gradient and resolution of symptomatology, regardless of prior surgical attempt at repair.

Recommendations

Given the low attendant morbidity of this procedure, we currently recommend BPV as the procedure of choice in adult patients with hemodynamically severe (peak systolic ejection gradient >80 mm Hg) or symptomatic moderate VPS (peak systolic ejection gradient >40 mm Hg and <80 mm Hg) either in

Figure 6: The technique of balloon pulmonary valvotomy.



the individual with unoperated VPS or with recurrent VPS after initial surgery. Particular caution to avoid prolonged reduction in systemic cardiac output should be taken in the elderly and in patients with severe VPS associated with low pulmonary cardiac output or right ventricular failure. The role of serial graded dilations in these circumstances has not been evaluated.

Coarctation of the aorta

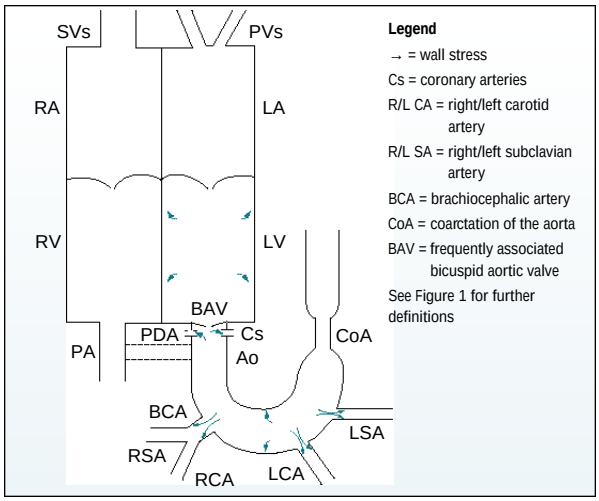
Clinically detectable coarctation of the aorta (CoA) in the adult (Figure 7), with a resting gradient ≥20 mm Hg between upper and lower extremities, carries increasing risk of progressive left ventricular dysfunction, persistent systemic arterial systolic hypertension, premature cerebrovascular and coronary atherosclerosis, and the potential for dissection or rupture of the aorta, coronary or cerebral vessels (especially during pregnancy, surgery, or catheterization).

The natural history of CoA (gradients ≤ 20 mm Hg) in the adult is undefined; however, modern surgical success is excellent with perioperative mortality ≤2% for native CoA repair. During long-term follow-up, re-coarctation may occur in 8% to 20% of patients in modern series, depending on the type of surgical repair. Modern surgical risk of mortality for re-operation is ≤2% with low risk of paraplegia and late aneurysm formation at the repair site.

In children, balloon dilation (BD) of recurrent or persistent CoA following surgical correction is now considered the therapy of choice and an effective alternative to surgical correction for native CoA.[48-53] Rates of success (defined as gradient reduction of ≥50% and an increase in angiographic luminal diameter ≥30%, or more recently, as a residual gradient ≤20 mm Hg) are over 80%. As well, morbidity is low when balloons that are three to four times the diameter of the CoA,(but less than 1-2 mm greater than the size of the normal aorta) are used. Our previous institutional bias favoring surgical repair versus BD of native CoA in the absence of severe left ventricular dysfunction due to improved gradient reduction has changed due to increasing use of balloon-assisted stenting of CoA.

Dilation of postoperative recurrent stenoses in 548 patients was performed in multiple centers in the Valvuloplasty and Angioplasty of Congenital Anomalies (VACA) Registry.[50,54] While the number of adults in this registry remains limited, there has been no detectable difference in risk of success or adversity based upon adult age. Less than a 20 mm Hg gradient was achieved in 75% of patients after BD. Procedure-

Figure 7: Aortic coarctation physiology. Narrowing in descending aorta,just distal to the origin of the left subclavian artery. Wall stress is raised in all proximal chambers and vessels, leading to potential for advanced atherosclerosis, vascular dissection or rupture, and chamber hypertrophy.



related death occurred in 0.7%, with peri-procedural stroke in 0.6%. Transmural (0.7%) or intimal dissection (1.6%) was noted, with rare need for immediate surgical repair. Along with the recent pooled VACA data demonstrating equivalent results of BD for native CoA and recurrent CoA, two additional series highlight similar results in single-center experiences with BD of native CoA in adolescents and adults (New Delhi (35 patients); Riyadh 43 patients).[52] Immediate success was achieved in 74% and 93%, respectively.

Balloon-assisted stenting of the aorta without precedent maximal balloon dilation permits use of smaller, non-"oversized" balloons, with less risk of dissection and rupture of the aortic wall.[55] Balloon-assisted stenting of native or recurrent CoA in 12 adolescents and adults at Boston's Children's Hospital has allowed for graded dilations and near-full relief of gradient, even in tubular stenotic areas.

Recommendations

We currently recommend balloon-assisted stent implantation or primary BD as the therapy of choice for all adults with recurrent CoA (defined as resting gradient ≥ 20 mm Hg), as well as in patients with native CoA and left ventricular dysfunction or those at significant risk from surgical repair due to medical co-morbidity. Sufficient data exists to recommend balloon-assisted stent implantation or primary BD as an effective alternative to surgical repair for all adults with native CoA. The role of stent implantation for relief of CoA gradient ≤20 mm Hg, or for women of childbearing age to decrease risk of subsequent rupture during pregnancy, has yet to be defined.

Proximal and peripheral pulmonary artery stenosis

In adults, balloon dilation and balloon-assisted stent implantation as primary therapy for native and postoperative narrowings in the pulmonary ventricular outflow, as well as in the proximal and distal pulmonary vasculature, have a similar success rate as in children. After surgical repair of tetralogy of Fallot, patients may have ≤25% incidence of recurrence or residual obstruction at any level of the right ventricular outflow tract. Balloon dilation or stent implantation remains the procedure of choice for the majority of such patients when

Table 1: Catheter-based interventions for the adult with congenital heart disease

Procedure of choice	Effective alternative to surgery	Unproven effect
Device closure	**Device closure**	**Device closure**
PDA	ASD-2	PFO (Ebstein disease)
VSD-postoperative residual	PFO (stroke)	
VSD-congenital muscular	VSD-post-MI	
Fenestrated Fontan baffle		
PFO (cyanosis)		
Balloon/stent dilation	**Balloon/stent dilation**	**Balloon/stent dilation**
Peripheral pulmonary stenoses	Native CoA	Subvalvar AS
Recurrent CoA	Conduit/baffle obstruction	TOF
		Pulmonary venous stenosis
Balloon valvulotomy	**Balloon valvotomy**	
Valvar PS	Valvar AS	
Coil embolization	**Coil embolization**	
Thoracic collaterals/PDA	Coronary artery fistulae	
Postoperative residual shunts	Pulmonary AVM	

additional surgery is not required and when obstruction is at the level of the pulmonary trunk or beyond. Extension of these techniques have led to balloon and stent expansion of surgical outflow tract conduits (Rastelli-type repair) including patients with homograft stenosis after the Ross (pulmonary autograft for aortic valve disease) procedure.

We have utilized similar techniques to assist 35 adults with either isolated peripheral pulmonary artery stenoses or acquired chronic distal thromboembolic pulmonary hypertension. Most were profoundly debilitated and were referred for evaluation for lung transplantation. The most frequently encountered complication was early development of transient "reperfusion pulmonary edema" in segments of lung with restored pulmonary blood flow after dilation. After three to four years of follow-up, survivors have improvement in exercise tolerance and have not required transplantation.

Systemic and pulmonary venous obstruction

Successful application of balloon dilation and balloon-assisted stent implantation as therapy for native, acquired, or postoperative narrowings in the systemic and pulmonary veins, such as after atrial switch repair or the Fontan procedure, has yielded similar success to that seen with children.

Conclusion

Recent ACC/AHA guidelines on interventional catheterization in patients with congenital heart disease underscore the "investigational" nature of most of the techniques outlined in this report.[56] However, these same guidelines recognize an inability to approach these illnesses with large-scale or randomized trials, and therefore, to advocate continued investigation and use of catheter-based diagnostics and therapeutics to support or supplant surgical procedures. Currently, we consider these techniques as procedures of choice, effective alternatives to surgical therapy, or treatments with unproven effects (Table 1).

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The Boston Adult Congenital Heart (BACH) Service is a multi-disciplinary and multi-institutional program designed to advance and coordinate the long-term care and investigation of young and older adults with congenital heart disease. Established as a regional center of expertise, BACH provides comprehensive care in all aspects of adult and pediatric cardiovascular and thoracic medicine, surgery, nursing, medical/pediatric sub-specialties, obstetrics/gynecology, genetics, psychiatry, exercise rehabilitation, and social services. Outpatient and inpatient care is offered at both BWH and CH with location of service dependent upon patient need and local expertise. Training of clinical and research fellows/visiting cardiovascular staff, occurs within both monthly and year-long programs. Similar programs are available for mid-level practitioners. BACH research interests and active protocols focus on pathogenesis and therapies of pulmonary vascular diseases, "atypical" systemic and right ventricular function, and novel transcatheter and surgical therapeutics for intracardiac/vascular shunting,stenoses and arrhythmias.

More information regarding programs and care available for adults with congenital heart disease through the Boston Adult Congenital Heart (BACH) Service can be obtained by calling the BACH office, 9 AM-5 PM, Monday-Friday, (617-355-6508: Tara O'Regan, administrative coordinator) or by paging the BACH physician-on-call (617-732-6660 beeper #3-BACH 32224).

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