The role of the aortic translocation in the management of complex TGA

—Nikaidoh Procedure—

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In 1980, Bex and associates (1) were the first to introduce the concept of aortic translocation for the management of transposition of the great arteries (TGA) but it was Nikaidoh (2), in 1984, who popularized the technique for the treatment of TGA with a ventricular septal defect (VSD) and pulmonary stenosis (PS). The repair consists of harvesting the aortic root from the right ventricle, relieving the left ventricular outflow tract obstruction by dividing the outlet septum and excising the pulmonary valve, reconstructing the left ventricular outflow tract (LVOT) with the translocated aortic root and the VSD patch, and the right ventricular outflow tract (RVOT) with a pericardial patch.

Modifications to the original technique include individual coronary transfer during translocation (to avoid the possibility of coronary ischemia), the use of the Lecompte maneuver, and RVOT reconstruction with a pulmonary homograft or direct right ventricle to pulmonary artery anastomosis. Although technically challenging, aortic translocation combines elements of commonly performed surgical techniques including the Ross, Konno and Jatene procedures.

The Nikaidoh procedure results in a more ‘normal’ anatomic repair; the right and left ventricular outflow tracts are better aligned, avoiding the right angle turns created by the Rastelli repair. Also, the right ventricle to pulmonary artery connection is less susceptible to sternal compression, which should decrease the incidence of reoperations for right ventricular outflow obstruction, especially when using a direct pulmonary artery to right ventricle connection (3).

In our experience, the only contraindication to the Nikaidoh procedure has been the presence of anomalous coronary anatomy. The epicardial course of a major coronary artery could preclude the safe harvesting of the aortic root from the right ventricle (i.e. right coronary artery originating from the left main). Also, the coronary anatomy could affect the ability to safely move the aorta posteriorly, into the pulmonary annulus (i.e. posterior intramural coronary artery course); this anatomy may also not be amenable to coronary reimplantation in conjunction with aortic translocation. In these patients the Rastelli or the REV procedures appear to be better options.

At Children’s Hospital of Pittsburgh the aortic translocation procedure is the preferred surgical technique for the management of patients with TGA with a VSD and PS. Although the overall “published
experience” [1-2,4-17] with this technique is limited, the early and midterm results are encouraging, suggesting improved outcomes when compared to the Rastelli repair. Certainly, in patients with ‘inadequate anatomy’ for a Rastelli or REV repairs the Nikaidoh procedure becomes an attractive alternative.

Bibliography


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Surgery for atrial fibrillation:
Finding our way out of the maze

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Following the success of the Cox-Maze procedure, multiple variations of the procedure recently have been performed on patients (1-6). Most of these procedures have involved the use of a smaller lesion set and have included only either right atrial Maze lesions, only left atrial Maze lesions, pulmonary vein isolation or pulmonary vein isolation with additional left and/or right atrial lesions. In general, all of these approaches have had lower success rates than the Cox-Maze III or IV procedures. Catheter-based approaches have demonstrated that in certain cohorts of patients, atrial fibrillation was initiated by triggers in the pulmonary veins and in some of these patients, was also maintained by a source in the pulmonary vein (7). Based on these developments, pulmonary vein isolation was applied to patients undergoing surgical treatment of atrial fibrillation. Unfortunately, mapping data have shown that the source of AF can be located outside the pulmonary veins in both paroxysmal and persistent atrial fibrillation (8, 9). This has been further demonstrated in studies showing that the failure rate of the Cox-Maze procedure increases as the left atrial size increases, independent of the type of atrial fibrillation (10, 11). Studies have shown poor results with pulmonary vein isolation alone in patients with long-standing atrial fibrillation (12).

Surgeons can look to what has been done in catheter based approaches in order to guide more patient-specific treatments of AF. First, patient selection should be considered. Most catheter based treatments are performed in patients with atria that are not enlarged (< 5.5 cm) and do not have concomitant disease (13). Most patients undergoing surgical treatment are undergoing other procedures, and often have enlarged atria. Caution should be exercised in concluding that what works in the nonsurgical patient population will work in the surgical patient population. Second, what cardiologists do to evaluate and test the patient before and after ablation should be taken into account. Prior to any catheter ablation, electrograms are recorded and used to evaluate local activation and tissue viability (14). These data are used to identify the source of the AF. Based on this information, ablations are performed to terminate the AF. If AF recurs, the process is repeated. In addition, following ablation that terminates the AF, attempts are made to reinitiate the AF using extrastimulation or drugs (15). This approach is radically different than a completely anatomically based treatment and allows for a more patient-specific treatment. This electrophysiologic information is essential if surgeons are to develop a patient-specific surgical ablation procedure. A third consideration is the end points of a procedure. Surgical and catheter based approaches must diverge here for both practical and ethical reasons. Surgical procedures have to be
designed to work the first time, whereas patients undergoing catheter treatments can undergo repeat procedures without a substantial increase in morbidity or mortality.

The Future of Surgical Treatment

Two advances are needed to facilitate a patient-specific treatment of AF. First, a noninvasive method of determining the source of a patient’s AF must be developed and validated. Despite the progress made on intraoperative mapping, it still remains difficult and time consuming, and most surgeons are not equipped to perform the mapping. A newer noninvasive technique, electrocardiographic imaging (16–18), offers a potentially useful technique for describing the atrial activation sequence and deriving mechanistic information. Electrocardiographic imaging applies an inverse solution to body surface electrograms recorded from 250 sites on the torso. The geometry is defined by a high resolution computed tomographic scan made with the electrodes, which contain radiographically opaque markers, in place on the torso. A three dimensional model of the heart, torso, and electrode geometry is created from the computed tomographic scan. Using the inverse solution to the potential field equations, potentials on the surface of the atria can be calculated from the torso surface potentials. The virtual atrial electrograms can then be analyzed to determine activation maps. These data can be obtained from conscious patients prior to surgery. We are testing this technique in collaboration with Dr. Yoram Rudy, the developer of electrocardiographic imaging, and the initial results are promising. The second advance needed is a strategy for developing a lesion set, based on the electrocardiographic imaging data, that terminates the patient’s AF, restores normal sinus rhythm and AV conduction, and minimizes the effect on atrial mechanical function. By using mechanistic information (refractory period and conduction velocity) derived from activation data and detailed anatomic data from the computed tomographic scan, the critical area needed to maintain AF can be calculated based on the principal of the critical mass hypothesis (19–21). The lesions of the Cox–Maze III procedure can be added or deleted based on these data. The goal is to create atria that cannot maintain AF. This goal is ambitious, but may allow for the development of minimally invasive treatments having high efficacy and safety.

References


Mitral valve repair is nowadays a recognized method to surgically treat mitral valve regurgitation. Basically, mitral valve regurgitation can be defined as a loss of an efficient surface of coaptation during systole. The aim of mitral valve repair is to restore a good surface of coaptation, thus restoring a competency to the mitral valve.

There is today little doubt that long time survival after surgery for mitral valve regurgitation is better after mitral valve repair than after mitral valve replacement. Two groups of patients operated in our institution with mitral valve regurgitation were compared. One group (433 patients) had mitral valve repair and the other (257 patients) mitral valve replacement with Medtronic–Hall prosthesis. The 7–year survival was 74% for the repair group and 58% for the replacement group. This difference was statistically significant. The durability over time is dependant of the etiology of the mitral disease as demonstrated by Carpentier’s team. A study recently published by this group showed at 25 years a 7% reoperation for the group of degenerative disease and an incidence of 53% of reoperation for the group of rheumatic disease. The trend nowadays is to operate patients with severe mitral valve regurgitation at an early stage when they are not yet symptomatic the goal being to preserve the left ventricular function that might deteriorate unnoticed.

In conclusion, improvements in surgical techniques, a better understanding in particular with the help of echocardiography have given mitral valve repair safety, predictability, and durability. Mitral valve repair can be proposed to non-symptomatic to prevent left ventricular dysfunction.

The tricuspid valve remains an enigma and tricuspid valve dysfunctions are too often ignored and untreated. Most of the time, tricuspid regurgitation is a finding in patients with advanced mitral valve disease. The tricuspid regurgitation is functional secondary to pulmonary hypertension and right ventricular dilatation leading to a dilatation of the tricuspid annulus. Nevertheless, it seems that dilatation of the tricuspid annulus should be treated even without regurgitation to prevent the onset of secondary tricuspid regurgitation despite successful left sided surgery.
How normal is normal left atrial anatomy in atrial fibrillation?

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Most researches into the mechanisms of atrial fibrillation (AF), in particular animal studies, convey the implicit message that the atria are structurally normal. This is somewhat surprising since the vast majority of patients with AF have associated cardiovascular diseases, such as hypertension and ischaemic and valvar heart disease. In fact several histo-pathological studies have shown extensive myocardial pathology in hearts of patients with AF. Moreover, atrial myocardium in humans shows structural changes that increase with aging, such as fibro-fatty changes and patchy replacement fibrosis, and AF is very much associated with older (> 35yrs) individuals. Why do these data receive little if any attention in considerations regarding the genesis of AF? It is of interest, therefore, that my own observations (Becker AE. Heart Rhythm, 2004; 1: 627-631) confirm previous works to this end and, in addition, suggest that these changes are more pronounced in hearts of patients with paroxysmal AF then in hearts of patients without AF.

The changes involve the myocardial sleeves that extend onto the pulmonary veins and sites of rapid conduction, such as the terminal crest and Bachmann’s bundle. Considering atria of patients with AF as structurally normal appears to be an illusion. In my opinion these structural changes should be taken into account as potential substrates for the initiation and maintenance of atrial arrhythmias in humans.
Management of the aorta in bicuspid aortic valve disease

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Aortopathy of bicuspid aortic valve disease is a topic of increasing interest and knowledge in cardiology and cardiac surgery. Bicuspid aortic valve disease is the most common congenital heart anomaly, affecting 0.9–2.0% of the population or approximately 4 million people in the US. It is the most common cause of aortic valve disease in patients under the age of 70. The preponderance of diseased bicuspid aortic valves suffer from stenosis (75%). Insufficient valves account for approximately 13% and mixed lesions 10%. Some bicuspid valves, however, an unknown percentage, reach adult life without any hemodynamically significant stenosis or insufficiency. Associations between BAV and abnormalities of the aorta dates at least to 1927, when it was noticed that aortic dissection appeared in patients with BAV. This was further developed in 1972 when McKusick noted the association between BAV and dissection of the ascending aorta with cystic medial necrosis. He favored a hemodynamic explanation. Lines of evidence began emerging in recent years about an association between BAV and proximal aortopathy. BAV is associated with other congenital cardiovascular lesions including coarctation, interrupted aortic arch and, more rarely, PDA, VSD and Turner’s or William’s syndrome. It has been noted that BAV increases the risk of dissection nine-fold in large registries. Furthermore dissections occur at a young age in BAV patients and usually occurs in the presence of a normally functioning BAV, contradicting the early impression that these aneurysms might represent post-stenotic dilation of the aorta beyond a stenotic BAV. Also emerging are anatomic data regarding aortic dilation in patients with BAV compared to normal aortic valves. Patients with BAV, irrespective or hemodynamics, have been found to have larger aortic dimensions at the Sinus of valsalva, the sinotubular junction and the proximal ascending aorta. This suggests a common developmental defect occurring in the aorta of patients with BAV. More recent data suggests that this aortopathy has distinctive patterns and usually involves the transverse aortic arch. The aorta in patients with BAV has been found to enlarge more rapidly than in patients with normal valves. Mechanistic studies have examined the extracellular matrix (ECM) and microfibrillar proteins as possible contributors to the aortopathy of BAV. Consistent patterns of deficiency of enzymes that destroy the matrix of the aortic wall have been found in patients with BAV. Furthermore, these increases in enzymes have been found to correlate with aortic diameter. Histologic studies as well as studies of apoptosis have demonstrated consistent abnormalities in the aorta of patients with BAV compared to the aorta of patients with a normal aortic valve.

Surgical implications of aortic pathology in bicuspid aortic valve are becoming better established. The
aortopathy typically involves the ascending aorta (sinotubular junction to proximal aortic arch), but may involve the aortic root as well as the aortic arch. Surgical studies in patients with BAV and aortic dilation undergoing aortic valve replacement, who have been followed for many years have been quite revealing. If an aorta greater than 4.5cm is left behind at the time of aortic valve replacement, the survival is statistically worse compared to patients having resection of enlarged aortas. Current recommendations for replacement are that an aorta of 4.5cm or greater at the time of aortic valve replacement in a patient with bicuspid aortic valve should undergo a replacement of the aortic segment that is enlarged, be it aortic root, ascending aorta and/or aortic arch. 2006 American College of Cardiology/American Heart Association guidelines for management of patients with aortopathy accompanying bicuspid aortic valve will be reviewed.
Stent grafts for aortic dissections: Future or folly?

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Thoracic stent grafts have become a new therapeutic modality, proven effective in the treatment of thoracic aortic aneurysms. Their utility for thoracic aortic dissections, however, remains more problematic and will be explored in this treatise.

Since the successful introduction by the Stanford group of stent grafts for the treatment of complicated type B aortic dissections, there has been significant progress and refinement in their utility. The work of Williams, et al at the University of Michigan, defining the mechanisms of malperfusion, significantly enhanced the utility of stent grafts for the treatment of dissections. Prior to the advent of thoracic stent grafts, open repair of dissected patients with visceral malperfusion resulted in surgical mortalities approaching 50%. With stent graft treatment, that mortality dropped to approximately 10%-15%. Additionally, reversal of the malperfusion could be monitored in the cath lab, with additional interventions at the branch vessel orifices to improve visceral perfusion.

Based on the efficacy in complicated dissections, enthusiasm grew for the application of stent grafts to uncomplicated acute type B dissections. This was eventually subjected to a randomized control trial, the INSTEAD trial, a multicenter European trial looking at stent graft treatment of uncomplicated type B dissections 14 days or more after the event, as compared to best medical therapy. The primary endpoint, aorta-related mortality at one year, resulted in a negative study, with optimal medical therapy resulting in a 97% survival and stent graft resulting in a 93% survival. However, secondary analysis at two years revealed essentially equivalent results when considering a combined endpoint of aortic-related death, the necessity for crossover or conversion, or other ancillary interventions. Many people, however, felt that this was a disadvantaged study, with most people surviving past day 14 without complications as having an exceptionally good prognosis, and does not really address the acute type B dissection.

Accordingly, the ADSORB trial, another prospective randomized multicenter European trial, has been organized, comparing stent graft therapy versus optimal medical therapy, randomized within seven days of presentation, with follow-up to three years of 250 subjects. That trial is currently enrolling patients.

In addition to conventional stent graft therapies, we have also seen the advent of new disease-specific devices: namely, the new Cook two-piece graft, a proximal-covered 10-cm segment with an uncovered 15
cm distally. This will accommodate an aortic diameter up to 40 mm and is specifically designed with low expansile strength to avoid tearing of fragile intima. This “PETTICOAT” concept has been preliminarily investigated in 29 patients at three institutions. Treatment success exceeds 90%. This has led to the STABLE trial, the study for thoracic aortic type B dissections using endoluminal repair. This trial will examine multiple endpoints, including branch vessel obstructions, resolution of the periaortic effusion/hematoma, resolution of refractory hypertension and pain, as well as looking at aortic enlargement at three and six months.

We also have more data concerning treatment of complicated type B aortic dissections. In 2002, Bufalo reported his initial 70 patients with complicated type B dissections, 35% of whom were acute. These sally results were quite appealing, with a 93% technical success rate and an acute mortality of 6%. In a more recent population, Bufalo now reports 106 patients with complicated type B aortic dissections treated with stent grafts, 45 in the acute phase and 61 in the chronic phase. There were 73 patients available for late follow-up, at a mean duration of 36 months. However, late failure was observed in 27 patients (37%), requiring reintervention in 16 patients, late surgical conversion in three patients, and death due to aortic causes in eight patients. Clearly, stent grafts may not be the long-term panacea we had hoped.

We have recently compiled data from six U.S. sites, utilizing the Gore TAG endoprosthesis for acute complicated type B dissections. We have compared this with the Parker, et al meta-analysis of 29 centers, comprising 942 patients. Early mortality is remarkably similar, 12.5% for the acute complicated dissections versus 9% for both acute and chronics with the Parker study, and a 4% paraplegia rate, compared to 2%. Looking at the contemporary results for open surgical repair in five recently published series, early results are comparable to endovascular management, with 15% mortality for open cases versus 12.5% for stent graft cases, and paraplegia rates 2.6% for open versus 3.8% for stent grafts. We also utilized this data, as well as the IRAD data, in an attempt to discern which patients warranted early elective stent graft treatment for both type A and type B acute aortic dissections. Two critical predictors of false lumen enlargement included an initial false lumen diameter greater than 22 mm, as well as a non-re-entry phenomena in the false lumen, producing partial false lumen thrombosis. Both of these predicted suboptimal 10-year survival.

Conclusion: Although stent grafts remain a fascinating modality for the treatment of aortic dissections, several areas of uncertainty remain. Hopefully, new evolving technology will be more user-friendly and new clinical trials will clarify indications and long-term results.

Clearly, there is still much to be learned in the treatment of this devastating disease, but progress is being made. Hopefully, we can continue to improve our understanding of these complex patients and their management, and evolve toward restoring these patients to a normal survival.
Surgical ventricular restoration: An operation to treat congestive heart failure in patients with dilated hearts after myocardial infarction.

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Early reperfusion of the infarct–related artery during myocardial infarction has become the standard of care. This can be achieved by thrombolysis or by percutaneous methods. In spite of early interventions, about 20% of patients develop ventricular dilation and further remodeling. This is often a progressive disorder leading to congestive heart failure.

A traditional surgical approach to this disease is resection of the left ventricular aneurysm if present. However, more often than not, a dyskinetic aneurysm is not seen because of early recanalization of the infarct–related coronary artery. Here is the ventricular wall is akinetic. Several years ago Dr. Vincent Dor developed an operation that is applied to both the dyskinetic or akinetic ventricle by placement of an intraventricular patch that excludes the scarred tissue, thereby reducing ventricular volume while reshaping the heart towards a more normal elliptical shape. This operation is one of several which can be collectively referred to as “ventricular restoration” or SVR.

The RESTORE Group consists of cardiologists and surgeons from 13 centers and four continents. Between 1998 and 2003 these investigators performed 1198 SVR operations. Concomitant procedures included coronary artery bypass grafting in 95%, mitral valve repair in 22%, and mitral valve replacement in 1%. Overall 30–day mortality after SVR was 5.3% (8.7% with mitral repair vs. 4.0% without repair, p < .001) Ejection fraction increased from 29.6 +/- 11.0% to 39.5 +/- 12.3% (p < .001) and left ventricular end systolic volume index decreased from 80.4 +/- 51.4 ml/m2 to 56.6 +/- 34.3 ml/m2 (p < .001). Overall 5–year survival was 68.6 +/- 2.8%. Logistic regression analysis identified EF < 30%, LVESVI > 80 ml/m2, advanced NYHA functional class, and age ≥ 75 years as risk factors for death. Five–year freedom from hospital readmission for CHF was 78%. Preoperatively, 67% of patients were class III or IV, and postoperatively 85% were class I or II. SVR improves ventricular function and is highly effective therapy in the treatment of ischemic cardiomyopathy with an excellent 5–year outcome.