Trends in vascular ring surgery

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Objective: We sought to review our experience with infants and children with anatomically complete vascular rings (ie, double aortic arch and right aortic arch with left ligamentum) and define perioperative trends in diagnostic imaging, operative techniques, and clinical outcomes.

Methods: From 1946 through 2003, 209 patients (113 with double aortic arch and 96 with right aortic arch) underwent surgical repair. Mean and median ages at the time of the operation were as follows: double aortic arch, 1.4 ± 2.4 years and 0.75 years, respectively; right aortic arch, 2.7 ± 3.9 years and 0.9 years, respectively. Fourteen (14.6%) patients with right aortic arch had an associated Kommerell diverticulum. Cardiac diagnoses were present in 26 (12.4%) of 209 patients.

Results: There has been no operative mortality since 1959. In the past 30 years, mean hospital stay decreased from 8 to 3 days. Primary means of diagnosis has shifted from barium swallow and angiography to computed tomographic scanning or magnetic resonance imaging. In the past 10 years, 73% of patients had preoperative or intraoperative bronchoscopy. The technique of operation has shifted to a muscle-sparing left thoracotomy without routine chest drainage. In 7 recent patients with right aortic arch and a Kommerell diverticulum, the diverticulum was resected, and the left subclavian artery was transferred to the left carotid artery as a primary procedure.

Conclusions: At our institution, computed tomographic scanning has replaced barium swallow as the diagnostic procedure of choice for vascular ring evaluation. We recommend both preoperative bronchoscopy and echocardiography. Use of a muscle-sparing thoracotomy without routine chest drainage has decreased mean hospital stay. For patients with a right aortic arch and associated Kommerell diverticulum, we recommend diverticulum resection with left subclavian artery transfer to the left carotid artery.

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The surgical series of infants and children undergoing division of a vascular ring at Children's Memorial Hospital was started by Willis J. Potts. He reported his experience with 2 cases of double aortic arch in 1948, the first of those patients undergoing surgical intervention on July 27, 1946.¹ This was only 1 year after the first surgical division of a vascular ring by Robert Gross at Boston Children's Hospital (June 9, 1945).² This tradition of being a referral center for these patients continued with the first major review of our experience by Drs Nikaidoh, Riker, and Idriss, which was published in 1972.³ This review included 33 patients with a double aortic arch and 26 patients with a right aortic arch and a left ligamentum. This experience was updated again in 1989 when we reviewed 61 patients with a double aortic arch and 52 with a right aortic arch and left ligamentum.⁴ Since then, the series has nearly doubled (Figure 1). The purpose of this current review is to evaluate perioperative trends in vascular ring operations over the past several decades, with particular attention to changes in diagnostic imaging, age at repair, improvements in surgical techniques, and ongoing evaluation of the

Figure 1. Vascular ring patient population, 1946-2004. Arrows indicate prior major publications from Children's Memorial Hospital.

clinical outcomes. In addition, we wanted to reevaluate and update the conclusions of our 1989 report.

Patients and Methods

The classification scheme of vascular rings used throughout this article is that recommended by the International Congenital Heart Surgery Nomenclature and Database Committee (Table 1).⁵ Our current review is limited to those patients with anatomically complete vascular rings (ie, double aortic arch or right aortic arch with left ligamentum). Patients with anatomically incomplete vascular rings (pulmonary artery sling and innominate artery compression syndrome) were excluded from this series.

From July 1946 through December 2003, 209 children have undergone surgical repair of a vascular ring at Children's Memorial Hospital. These patients were identified with our database, and their charts were reviewed. Institutional review board approval for chart review was obtained. This includes 113 patients with a double aortic arch and 96 patients with a right aortic arch and left ligamentum. Mean and median ages at the time of the operation were as follows: double aortic arch, 1.4 ± 2.4 years and 0.75 years, respectively; right aortic arch, 2.7 ± 3.9 years and 0.9 years, respectively. Sex distributions were 1.3:1 and 1.8:1 (male/female) in the double aortic arch and right aortic arch groups, respectively. Fourteen patients with a right aortic arch had a significant associ-

TABLE 1. Vascular ring hierarchy as defined by the International Congenital Heart Surgery Nomenclature and Database Committee⁵

- Double aortic arch
 - Right arch dominant
 - Left arch dominant
 - Balanced arches
- Right aortic arch–left ligamentum
 - Mirror-image branching
 - Retroesophageal left subclavian artery
 - Circumflex aorta
- Innominate artery compression
- Pulmonary artery sling

Figure 2. The anatomy of a patient with a right aortic arch, retroesophageal left subclavian artery, and large Kommerell diverticulum. The ligamentum is not illustrated. The Kommerell diverticulum is an embryologic remnant of the left fourth aortic arch. The diameter of the Kommerell diverticulum is usually equal to the size of the descending aorta. LCA, Left carotid artery; RCA, right carotid artery; LSA, left subclavian artery; RSA, right subclavian artery. Reprinted with permission of the artist, R. F. Idriss.

ated Kommerell diverticulum (Figure 2).⁶ Associated cardiac diagnoses were present in 26 (12.4%) of 209 of all patients with vascular rings, 18 patients with a right aortic arch (18/96, 19%), and 8 with a double aortic arch (8/113, 7%; Table 2). Other

TABLE 2. Associated cardiac diagnoses

Right aortic arch (n = 96)	
VSD	8
ToF	3
PDA	3
Congenitally corrected TGA	2
Absent LPA	1
Dextrocardia	1
Total	18
Double aortic arch (n = 113)	
Left SVC	2
Pulmonary atresia	1
Congenitally corrected TGA	1
VSD	1
PDA	1
Absent LPA	1
Dextrocardia	1
Total	8

VSD, Ventricular septal defect; ToF, tetralogy of Fallot; PDA, patent ductus arteriosus; TGA, transposition of the great arteries; LPA, left pulmonary artery; SVC, superior vena cava.

associated medical conditions include cleft lip-palate (n = 1), subglottic stenosis (n = 1), and complete tracheal rings (n = 1) in the double aortic arch group and Down syndrome (n = 4), subglottic stenosis (n = 1), tracheal hemangioma (n = 1), tracheo-esophageal fistula (n = 1), plastic foreign body in bronchus (n = 1), esophageal cyst (n = 1), scoliosis (n = 1), and end-stage renal disease (n = 1) in the right aortic arch group.

Clinical Presentation

The most common clinical symptom of these patients with an anatomically complete vascular ring was stridor (noisy breathing). A brassy cough, similar to a “seal bark,” was particularly common in the patients with a double aortic arch. Many patients had recurrent upper respiratory tract infections, chronic cough, or both. Dysphagia for solid foods was noted only in older children. The number of patients presenting with each of the above symptoms is shown in Table 3. Two patients presented with a coin lodged in the thoracic esophagus, and 3 others had vascular rings diagnosed at the time of removal of foreign bodies (chicken, chicken bone, and unnamed, respectively) from the esophagus. Several patients had either apneic or cyanotic episodes. Findings at the time of physical examination included rhonchi, stridor, retractions, wheezing, tachypnea, and rales. Some infants had opisthotonic posturing with hyperextension of the head to relieve the tracheal compression. In general, the patients with a double aortic arch had earlier onset of symptoms than the patients with a right aortic arch and left ligamentum.

Diagnostic Imaging

A chest radiograph was obtained in all patients, which frequently showed abnormalities. In particular, varying degrees of atelectasis, pneumonic infiltrates, and hyperaeration were common. The location of the aortic arch in relation to the trachea in many cases was helpful in determining the type of vascular ring. In the early time period of this study, barium esophagram was often the only other diagnostic procedure performed before surgical intervention. Angiography was also used in the early part of this series. More recently, computed tomography (CT) and magnetic resonance imaging (MRI) are commonly performed, even before surgical consultation. As bronchoscopy has become more widely applied, a growing number of patients have undergone bronchoscopy either preoperatively or at the time of vascular ring division. The number of patients having each of these different studies was compared and evaluated for trends over time.

Operative Findings and Management

Double aortic arch. In the majority of patients, the approach to a double aortic arch was through a left thoracotomy (108 patients). In 2 more recent patients, because of anatomic findings on the CT scan, the approach was through a right thoracotomy. One patient had congenitally corrected transposition of the great arteries, a dominant left aortic arch, and a right-sided descending aorta. The other patient had a dominant left aortic arch, an atretic right aortic arch, and an absent left pulmonary artery.

In the past 10 years, all patients approached through a left thoracotomy have had a muscle-sparing approach. The lung is retracted, and the pleura overlying the descending aorta is opened. The ligamentum arteriosum, left arch, right arch, left subclavian artery, left carotid artery, and descending thoracic aorta are all

TABLE 3. Symptoms leading to clinical presentation in patients with vascular rings*

	Double aortic arch (n = 80)†	Right aortic arch (n = 78)†
Stridor	46 (57%)	18 (23%)
Recurrent upper respiratory tract infections	22 (27%)	18 (23%)
Cough	17 (21%)	8 (10%)
Dysphagia	12 (15%)	12 (15%)
Respiratory distress	8 (10%)	13 (17%)
Ventilator preoperatively	7 (9%)	3 (4%)

*More than one symptom occurred in many patients. †Our records did not provide symptoms for the earlier patients in the series.

clearly identified. The vascular ring caused by the double arch is released by dividing the lesser of the 2 arches. If there is an atretic portion of an arch, this is the ideal site for arch division. The arches were divided between vascular clamps (Potts or Castañeda) and oversewn with fine polypropylene sutures (5-0 or 6-0). Pulse oximetry, palpation, or both of the pulses by the anesthesiologist was used to monitor the upper extremity and carotid pulses, which were checked after vascular clamps were applied and before arch division. In patients with balanced arches, the lower extremity blood pressure was checked, alternately occluding the right and left arch. The arch that led to a lower blood pressure with a test clamp was divided. If pressure measurements showed no change, the right arch was divided because the right arch wraps around the trachea and has higher potential to still cause tracheal compression. The left ligamentum was divided in most instances. With a dominant left arch, the ligamentum is of no consequence and does not necessarily require division. If the right arch is dominant, dividing the left arch but leaving the ligamentum intact will leave the patient with a vascular ring.

The recurrent laryngeal and phrenic nerves were identified and protected during the operation. An important final maneuver is to divide any and all fibrous bands adjacent to the esophagus and trachea. The pleura is always left open. In the past 5 years, more than 70% of patients were managed without use of a tube thoracostomy postoperatively. The pleural space was evacuated with a small suction catheter passed through the incision, and the catheter was removed at the time of wound closure.

Right aortic arch and left ligamentum. A left thoracotomy was used in 84 patients, and a median sternotomy was used in 12 (4/12 early in the series for unknown reasons). No patients were operated on through a right thoracotomy. Seven of the 8 more recent patients who had a median sternotomy approach also had an associated intracardiac repair. One previously reported patient⁷ had a right aortic arch, right ligamentum, and absent left pulmonary artery. The right ligamentum was divided through a median sternotomy, and the pulmonary artery was pexed to the aorta. In the usual case the vascular ring is released by dividing the ductus arteriosus or ligamentum arteriosum and carefully dissecting the trachea and esophagus free of adhesive bands.

Our recent experience with reoperation for recurrence of symptoms in patients with a Kommerell diverticulum who had simple ligamentum division has emphasized the importance of the asso-

Figure 3. Resection of a Kommerell diverticulum through a left thoracotomy. There is a vascular clamp partially occluding the descending thoracic aorta at the origin of the Kommerell diverticulum. The clamp on the left subclavian artery is not illustrated. The Kommerell diverticulum has been completely resected. *LCA*, Left carotid artery; *RCA*, right carotid artery; *LSA*, left subclavian artery; *RSA*, right subclavian artery. Reprinted with permission of the artist, R. F. Idriss.

Figure 4. The completed repair. The orifice at which the Kommerell diverticulum was resected is usually closed primarily, or as shown in the *inset*, the orifice can be patched with polytetrafluoroethylene if necessary. The left subclavian artery has been implanted into the side of the left common carotid artery with fine running polypropylene sutures. *LCA*, Left carotid artery; *RCA*, right carotid artery; *LSA*, left subclavian artery; *RSA*, right subclavian artery. Reprinted with permission of the artist, R. F. Idriss.

ciated Kommerell diverticulum.⁸ Earlier in the series, if the patient had a Kommerell diverticulum, it was pexed to the spine posteriorly. In one case a portion of the diverticulum was excised. In the recent time period (the past 5 years), the ligamentum is divided, the Kommerell diverticulum is resected, and the left subclavian artery is transferred to the left carotid artery. After dissection of these structures, the patient is anticoagulated with 1 mg/kg heparin administered intravenously. The base of the Kommerell diverticulum and a portion of the descending aorta are occluded with a Satinsky clamp (Figure 3), and the distal subclavian artery is occluded with a Castañeda clamp. The Kommerell diverticulum is resected. The base of the Kommerell diverticulum is oversewn with 2 layers of polypropylene (Figure 4). Alternatively (Figure 4, inset), the aorta can be patched with a small piece of polytetrafluoroethylene, particularly if the base of the Kommerell diverticulum is friable. The left subclavian artery is then anastomosed to an opening created in the left carotid artery, which is temporarily occluded with a Castañeda clamp. Patients were evaluated for the postoperative length of stay, significant postoperative complications, and late reoperation.

Results

There has been no operative mortality after repair of a vascular ring since 1959. Mean and median hospital stays have decreased over time (Figure 5). For the overall group, the mean and median hospital stay for double aortic arch was 6.9 ± 17.1 and 3 days, respectively, and for the right

aortic arch, these values were 5.8 ± 6.1 and 4 days, respectively. In the past 5 years, the mean hospital stays for double aortic arch and right aortic arch were 2.5 and 3 days, respectively.

In patients with a double aortic arch, the right-sided (posterior) arch was dominant in 85 (75%) patients, the left-sided (anterior) arch was dominant in 20 (18%) patients, and the arches were equal in size in 8 (7%) patients. With a

Figure 5. Mean postoperative length of stay after vascular ring surgery.

dominant right aortic arch, segments of the left arch were atretic in 34.1% (29/85) of the patients. With a dominant left aortic arch, segments of the right arch were atretic in 25% (5/20) of the patients. The atretic area occurred almost invariably in the posterior or distal end of the lesser arch.

There were 2 major anatomic variations of right aortic arch. In 66% (63/96) of patients, a retroesophageal left subclavian artery was present. In 34% of patients, there was a mirror image left innominate artery. In both situations, the vascular ring was completed by the ligamentum arteriosum, which stretched from the descending aorta to the pulmonary artery. This frequency of distribution is similar to that reported by Felson and Palayew (56% retroesophageal and 44% mirror-image branching).⁹

Morbidity was chiefly related to airway issues (tracheomalacia and bronchomalacia), prolonging postoperative stay. There were no reoperations for bleeding and no wound infections. One patient required urgent reoperation 1 day postoperatively to oversee a bleb leaking air from the left lung. Four patients (2% of the total series), 2 with double aortic arch and 2 with right aortic arch and left ligamentum, required late reoperation for aortopexy at 2 months, 5 months, 6 months, and 4 years, respectively, after the initial vascular ring division. The indication for reoperation in all cases was recurrence of airway symptoms.

The primary means of diagnosis in the past decade at our institution has shifted from barium swallow and angiography to CT scan or MRI (Figure 6). Until 1991, 66% of patients had either a barium swallow or angiography. Since 1992, that incidence has decreased to 44%. In contrast, the use of CT scanning or MRI has increased in the same time period from 29% to 80%.

Many patients were initially identified by the otolaryngology service at the time of bronchoscopy in the workup of the child with wheezing or stridor (Figure 6). In the past 10 years, 73% of patients have had a bronchoscopy as part of their evaluation. Bronchoscopy allows an assessment of the degree of tracheal compression, amount of tracheomalacia and bronchomalacia, and additional tracheal pathology. Three percent of these patients had such unanticipated findings. There were no iatrogenic complications related to bronchoscopy.

Our operative approach has been changed by the use of routine muscle-sparing thoracotomy incision and avoidance of routine chest drainage. Neither technique was used in the late 1970s. All patients now have a muscle-sparing thoracotomy, and most have no postoperative chest tube (32 patients over the past 10 years). None of our patients has required emergency placement of a thoracostomy tube for pneumothorax, hemothorax, or postoperative pleural effusion, as identified by means of postoperative chest radiography.

Figure 6. Diagnostic imaging techniques for the preoperative evaluation of vascular rings. There has been a dramatic shift from barium swallow and angiography to CT and MRI. There has also been an increase in the number of patients having preoperative or intraoperative bronchoscopy.

Our successful experience with reoperation for patients with a Kommerell diverticulum led us to recommend this as a primary operation. This procedure, resection of the Kommerell diverticulum and transfer of the left subclavian artery to the left carotid artery, has now been performed successfully as a primary operation in 7 patients. Age at surgical intervention in this subgroup ranged from 1.5 to 16.4 years (mean, 9.0 years). Mean left carotid artery clamp time was 19 minutes. Hospital stay in this subgroup ranged from 3 to 7 days (mean, 4.7 days). Histologic evaluation of the resected specimens has revealed medial necrosis. All have a patent left carotid–left subclavian artery anastomosis, as assessed by means of physical examination of the left radial pulse and echocardiography.

Discussion

The surgical care of children with vascular rings has evolved and matured over the decades.¹⁻⁵ The early work focused on elucidating the various congenital anatomic causes of respiratory distress and the basic surgical principles to relieve these anatomic problems. Over the past 2 decades, the focus has been on increasing the accuracy of the preoperative diagnosis and improving the surgical techniques to minimize postoperative morbidity without compromising patient safety. This analysis of more than 200 patients with true vascular rings (anatomically complete)—double aortic arch and right aortic arch with left ligamentum—has been accumulated over nearly a 60-year time period. The paradigm shifts in the care of these patients over the past decade have primarily been in terms of techniques of diagnosis, reduced surgical morbidity, decreased length of stay, and, for one diagnosis, an initial operation of greater magnitude to prevent later reoperation.

There has been a major shift in our strategy of preoperative evaluation of these patients since our most recent

review of this series.⁴ In 1989, we concluded that barium esophagram was the single most important study for the evaluation of patients with a vascular ring. This was also the recommended primary diagnostic study at other centers.¹⁰ Because of the increasing ease and safety of CT and MRI and the improvement of patient outcome achieved by the more precise ability to plan the operative procedure, we no longer believe that a barium esophagram is sufficient before proceeding with vascular ring repair. We now obtain a preoperative CT or MRI in all patients with vascular rings. Our preference for CT (over MRI) is based on the speed at which the scan can be obtained in a child with a marginal airway on the basis of the multidetector technology.¹¹ Multidetector technology has increased the speed of CT scans, and they can now be obtained in 10 to 20 seconds. This compares with 30 to 45 minutes required for a sedated MRI. The CT scan is relatively insensitive to motion and metal artifact in contrast to MRI. Reconfiguration of the CT scan as a 3-dimensional image is particularly useful.¹² Within the past 10 years, we have had several patients in whom a left thoracotomy approach (the standard approach used when the barium swallow shows a vascular ring) would not have been anatomically appropriate. There are a number of reviews that have analyzed these rare patients who require a right thoracotomy for optimal surgical exposure and results.¹³⁻¹⁵ I have had personal communication with surgeons who have had to perform 2 separate thoracotomies to divide a vascular ring when they relied solely on the barium swallow and echocardiogram for the diagnosis. These lesions can all be identified with a preoperative CT or MRI, which provide a clear surgical road map allowing a precise preoperative strategy.

We now recommend a preoperative or intraoperative bronchoscopy for all patients with vascular rings. We believe that a blind intubation in these patients is more dangerous than a controlled bronchoscopic evaluation to assess proper endotracheal tube position and the tracheal pathology that has to be managed. Three percent of the patients in our series had additional tracheal pathology not noted on other studies. These patients frequently have tracheomalacia, bronchomalacia, or both caused by the vascular ring.^{16,17} This becomes important when talking with the family with regard to the patient's short- and long-term prognosis.

In our review the overall incidence of congenital heart disease in patients with vascular rings was 12%. Because of this association, we currently recommend a preoperative echocardiogram in all patients with vascular rings for the purpose of identifying congenital cardiac lesions and not for vascular ring anatomy. This is also a change from our recommendations in 1989. At one point, we and others were hopeful that echocardiography with Doppler color flow imaging would be useful in helping to evaluate the actual

anatomy of the aortic arch anomaly. However, because of the limited windows available to the echocardiographer and because vascular ring segments without lumens cannot be displayed, echocardiography has not become a diagnostic tool for defining arch anatomy.

In the 1990s, our service was referred a number of patients who underwent operations elsewhere for repair of a vascular ring resulting from a right aortic arch and a left ligamentum and had a Kommerell diverticulum that was simply observed at the time of ligamentum division. This group of patients presented with recurrence of their respiratory symptoms, dysphagia, or both after successful ligamentum division. The Kommerell diverticulum in each of these individuals was an independent cause of tracheoesophageal compression. We reported successful reoperation on 8 such patients⁸ and have now successfully performed reoperations on 10 patients. The surgical procedure used was to resect the Kommerell diverticulum and reimplant the left subclavian artery into the left carotid artery. Histologic evaluation of the resected specimens revealed medial necrosis, which places the patient at risk for late aneurysm formation.

Because of this experience, we began to recommend resection of the diverticulum as a primary operation. The anastomosis of the left carotid artery to the left subclavian artery essentially reduplicates the normal innominate artery configuration. We have now performed this procedure successfully as a primary operation in 7 patients. Langlois and colleagues¹⁸ (vascular ring series of more than 500 patients) also recommend this, stating, "If it exists, the Kommerell's diverticulum must be divided between two clamps. The left subclavian artery must be divided at the level of the aorta and then reimplanted into the left common carotid artery." I have had personal communications with other surgeons who have successfully treated patients with Kommerell diverticulum resection after unsuccessful simple ligamentum division (Jonas R, personal communication, March 2003).

The other surgical techniques that we have modified over the past decade have shortened the postoperative morbidity and hospital stay of our patients. We adopted routine use of a muscle-sparing incision once we realized that these muscles can be easily mobilized without division and without any sacrifice of the degree of surgical visibility necessary at the time of the procedure. This results in less chest wall discomfort and improved patient mobility and arm mobility, facilitating the postoperative recovery. After noting for many years that most patients drained almost nothing from their tube thoracostomy in the postoperative period, we began routinely not placing a chest tube at the time of the operation (except in patients with Kommerell diverticulum). By avoiding placement of a chest tube and using early

extubation, many of the patients with routine vascular rings can be discharged at 36 to 48 hours postoperatively.

Conclusion

At Children's Memorial Hospital, our series of patients with anatomically complete vascular rings (double aortic arch, right aortic arch, and left ligamentum) now numbers more than 200. The most common presenting symptoms were stridor, recurrent upper respiratory infection, cough, and dysphagia. Review of trends in our management strategy revealed that CT scanning with contrast has replaced barium swallow as the diagnostic procedure of choice. We recommend preoperative or intraoperative bronchoscopy to rule out associated intratracheal pathology and provide an assessment of bronchomalacia-tracheomalacia. We also recommend a preoperative echocardiogram to rule out cardiac pathology, which was present in 12% of our patients. For patients with a right aortic arch and an associated Kommerell diverticulum, we recommend Kommerell diverticulum resection with left subclavian artery transfer to the left carotid artery. This should prevent the need for late reoperation in this subgroup of patients. The use of a muscle-sparing thoracotomy incision without routine chest tube placement has improved the duration of hospital stay. Patients with vascular rings require a high index of suspicion to come to the diagnosis, but once the diagnosis is established, surgical therapy can be accomplished with very low morbidity and essentially no mortality.

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Discussion

Dr Mark W. Turrentine (Indianapolis, Ind). Dr Backer and his colleagues continue to advance the understanding and management of vascular ring anomalies with their extensive history at Children's. This updated collection of cases reviews the evolution of their experience, including the integration of advanced diagnostic modalities, surgical approaches, and management of associated anomalies. On the basis of the addition of this recent information, the authors redefine the contemporary management of patients with this collection of defects.

Two specific areas of questions emerged from your conclusions, Carl. In your article you note that the majority of your patients are referred through the otolaryngology service and, in large part, have a preexisting bronchoscopy, which yields 6 patients, or about 3% of your series, with associated tracheal pathology. These pathologies include subglottic stenosis, transesophageal fistula, and complete cartilaginous rings. You then follow this with either a CT scan or an MRI for definitive diagnosis and reserve echocardiography for identification of concomitant cardiac lesions.

At our institution, most of our patients arrive with a preexisting barium swallow and chest radiogram. We then proceed with an echocardiogram, and in the vast majority of cases, this definitively identifies the double aortic arch or right aortic arch with retroesophageal subclavian. And it has been very good at telling us which arch is dominant. With rare exception, we have added a CT scan and, more recently, an MRI to make the definitive diagnosis.

In this era of cost-effective medicine, can we reconcile the routine addition of both bronchoscopy and, more importantly, these other expensive and albeit state-of-the-art diagnostic modalities to that of perhaps a preexisting barium swallow and echocardiogram as required diagnostic modalities when there is a seemingly low yield of pathology found by these studies requiring intervention at the same setting, or can we selectively apply these modalities and maintain an acceptable standard of care?

Dr Backer. Those are very good questions, Mark.

In general, we have been disappointed with the ability of the echocardiographers to give us an accurate preoperative anatomic

assessment. When you compare an echocardiogram with the detail that you get on a CT scan, it is very difficult to go back to the old days of operating with just a barium swallow and an echocardiogram.

There are many useful anatomic details to be learned from the CT scan. One would be identifying a Kommerell diverticulum. Another is in a patient with balanced aortic arches, in whom it is helpful to know which arch is larger and also whether the approach should be a right or a left thoracotomy. That is information that, in my opinion, is not always obtainable from the echocardiogram.

Dr Turrentine. Regarding your approach to the Kommerell diverticulum, presumably many of these diverticula have been approached in the past with aortic plication and aortopexy and have been successfully managed. We have not experienced a return of any of these patients to Riley Hospital, but I am aware of one adult patient in the community who was recognized by a colleague.

My question, regarding the diverticulum, is as follows: what criteria do you use to actually define the characteristics and make the diagnosis of a diverticulum?

Dr Backer. Most of these diverticula are actually about the same size, approximately the size of the descending thoracic aorta. If you looked at the CT scan of the smallest child that I operated on, the diverticulum size in relation to the aorta looks proportionally the same as in the oldest child in the series with a diverticulum. I do not think this diverticulum is enlarging over the childhood years but rather is a relatively stable congenital anomaly that independently compresses the trachea and the esophagus.

Dr Turrentine. Were you able to tease out any specific characteristics in those patients referred to you for symptomatic recurrence that might help identify those patients at the primary operation as being prone to redevelopment of symptoms?

Dr Backer. Most of the patients who underwent reoperation for a Kommerell diverticulum had a recurrence of their symptoms that temporarily went away after the original ligamentum division. That is why we switched to electively resecting the Kommerell diverticulum as a primary operation, to prevent recurrence of symptoms.

Dr Turrentine. Finally, do you have any histologic information on the basis of those specimens that you resected that might impart some knowledge as to whether this is abnormal aortic wall tissue, such as cystic medial necrosis, that might also validate the need to routinely excise this tissue?

Dr Backer. We actually do have the histologic specimens. We have not specifically looked at them, but it is an excellent idea to go back and examine these carefully and see whether they have cystic medial necrosis or some other pathologic problem.

Dr John E. Foker (Minneapolis, Minn). Very nice presentation of what is obviously the definitive series. You are to be commended on trying to improve the operation and shorten the hospital course. I have one question with regard to the operative procedure.

When I operate on these children, I find that the esophagus lies lateral to the trachea and not behind it. The esophagus can be mobilized for much of the length of the thoracic cavity, which provides more room for the trachea. The diverticula do not seem to be an issue; they lie well lateral to the trachea, and with the esophagus extensively freed up, it is not susceptible to obstruction.

Why do you believe it is necessary to remove these diverticula that do not seem to produce difficulties?

Dr Backer. The diverticulum, if left in place, most commonly causes recurrence of the respiratory symptoms and less commonly causes recurrence of swallowing symptoms. I have had 5 patients of those 10 reoperations who had recurrent dysphagia. When I reoperated on them, the diverticulum was clearly pushing on their esophagus.

The main reason for reoperation in these patients is independent compression of the trachea by the Kommerell diverticulum. One of these patients had the ligamentum divided and initially did well. Then, over the course of the following years, every winter she had 10 or 20 colds that she could not seem to shake. The winter after I resected her Kommerell diverticulum, she was free of events.

Dr Gerhard Ziemer (Tuebingen, Germany). Well, I can support Dr Backer's last notion because we operated on our most recent patient this January just for this diverticulum, which had not been resected 12 years before when the vascular ring was divided.

But when it comes to simplifying the procedure, our approach for resection of a Kommerell diverticulum is almost always from the side of the descending aorta, most often through a right-sided thoracotomy. Although you seemed not to have any problems with your routine left-sided approach, I agree with Dr Foker's preference to avoid digging deep behind the esophagus. I would be worried to think of a descending aortic bleeding site in the opposite side of the chest. Maybe you never have seen any bleeding in Chicago, but I prefer direct access to the descending aorta from the other side.

Dr Backer. We do not have bleeding in Chicago, no!

I would say, though, can I ask you before you leave the microphone, what do you then do with the left subclavian artery? We have reimplemented the left subclavian artery. It is one thing to ligate and divide the left subclavian artery in a newborn, but in someone who is 15 or 16 years old, I would not want to divide the subclavian artery without reimplanting it.

Dr Ziemer. Just talking about the patient from this January, she was 38 years old, 12 years after the primary operation through a left-sided thoracotomy. After resecting the Kommerell diverticulum through a right-sided thoracotomy and thereby dividing the subclavian artery, we looked for the pulse on the left radial artery, and it was diminished but sufficient. Therefore we let it go. I have to admit that this is not a corrective operation, but I feel myself and the patient being on the safe side of life.

Dr Joseph J. Amato (Chicago, Ill). Carl, this was an excellent presentation. The discussions seem to be focusing on the Kommerell diverticulum.

I can recall some 35 years ago operating on a 225-pound policeman with a vascular ring. For all of his life, as he remembers, he swallowed his steaks by chewing them first and then lifting his chin up and letting the steak go by the constriction of the ring and then lowering his chin. After dividing the ring, I had to reoperate on him, even though I had dissected the loose tissues to free the aorta from the esophagus. The diverticulum had continued to compress his esophagus. I folded the diverticulum on itself. Since then, I have always imbricated the diverticulum away from the esophagus, and I believe that is an important step in the operation. I would like your comments on it.

Second, I would also like your comments on the reported articles that describe the methods of doing vascular rings through a thoracoscope.

Dr Backer. Thanks for your comments, Dr Amato.

For the first patient in that reoperation article, I actually resected the diverticulum without transferring the subclavian artery. That patient continued to have mild symptoms from the sling-like effect of the left subclavian artery pulling on the aorta. After that experience, every other patient in our reoperation series underwent division and transfer of the left subclavian artery to the left carotid artery. It is a relatively simple operation, and it completely relieves that posterior compression.

The issue of thoracoscopic division is a very appropriate one, and there is some very good work going on regarding that. Our main issue with thoracoscopy has been the risk of putting a clip on these arches, of which 75% (in our series) are patent. As someone mentioned, the arch, when divided, retracts back into the mediastinum. If that clip should slip off with the arch retracted and not visible, it would be a very difficult situation to recover. Nevertheless, there have been thoracoscopic and robotic divisions of vascular rings reported.

Dr Smruti Ranjan Mohanty (Bangalore, India). I have a question for you. For this follow-up of this patient in the late phase, have you noticed any gradient across the arch?

Dr Backer. Any gradient across the anastomosis?

Dr Mohanty. Not anastomosis.

Dr Backer. Of the descending thoracic aorta?

Dr Mohanty. Not even descending. In the arch, the residual arch.

Dr Backer. Oh, I see what you are asking. Have we had a coarctation develop in the patent arch?

Dr Mohanty. Yes.

Dr Backer. We have not. I know the San Francisco group has reported a relatively high incidence of that occurrence. I have been curious enough to look for this in our own patients with a double arch, especially those patients with equally sized arches. We are always careful to assess the blood pressure in the leg during the operation. I have thought, in the last couple of years, that I am going to eventually run into a patient with a stenotic arch. If that occurs, we are of course prepared to do a coarctation repair at the same time, but we have not had that experience in our series.

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