

Accessory and Anomalous Atrioventricular Valvar Tissue Causing Outflow Tract Obstruction

Surgical Implications of a Heterogeneous and Complex Problem

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Objectives. The purpose of this study was to determine the effect of accessory or anomalous atrioventricular valvar apparatus on relief of outflow tract obstruction.

Background. Outflow tract obstruction due to accessory tissue or anomalous attachments of the atrioventricular valvar apparatus is an unusual but well-recognized problem. In addition to obstruction, anomalous attachments of the atrioventricular valvar apparatus may interfere with procedures to relieve outflow tract obstruction or perform outflow tract reconstruction.

Methods. Since 1992, we have operated on 21 patients (median age 4 years) with systemic (n = 13), pulmonary (n = 5) or bilateral (n = 3) outflow tract obstruction due to accessory atrioventricular valvar tissue and/or anomalous attachments of the subvalvar apparatus. Primary diagnoses were isolated obstruction of the systemic outflow tract or aortic arch (n = 7), transposition complexes (n = 6), previously repaired atrioventricular septal defect (n = 3), functionally single ventricle (n = 3) and ventricular septal defect with pulmonary outflow obstruction (n = 2). Outflow tract gradients ranged from 20–110 mm Hg (median 58 mm Hg).

Results. Complete relief of obstruction due to atrioventricular

valvar anomalies was possible in 14 patients. In six patients, the planned procedure either had to be modified or only partial relief of the obstruction was achieved. In the remaining patient, who had borderline functionally single ventricle heart disease (unbalanced atrioventricular septal defect) and systemic outflow obstruction due to accessory and functional valvar apparatus, support was withdrawn because the parents refused univentricular palliation and the valvar anomalies precluded a Ross-Konno procedure. There were two early deaths. At follow-up ranging from 1 to 66 months (median 27 months), there was one death, and there has been no recurrence of outflow tract obstruction or residual atrioventricular valvar tissue.

Conclusions. Outflow tract obstruction caused by accessory or anomalous atrioventricular valvar structures is an uncommon and heterogeneous group of conditions that can have significant surgical implications. In the majority of cases, tailoring of surgical techniques will permit complete relief of obstruction. However, such anomalies may limit standard surgical options and necessitate an innovative approach in some patients.

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Outflow tract obstruction due to accessory atrioventricular (AV) valvar tissue or anomalous attachments of the tensor apparatus is an uncommon problem (1–21). In addition to causing obstruction per se, accessory tissue or anomalous attachments of either AV valvar apparatus may interfere with procedures to relieve outflow tract obstruction or perform outflow tract reconstruction in patients with congenital heart disease. Such anomalies may be isolated or found with a wide range of associated lesions, including simple fibromuscular subaortic obstruction, AV septal defects, transposition complexes, and hearts with a functionally single ventricle among others. Although these phenomena have been described in a

number of autopsy and surgical cases, such reports have generally consisted of only one or two patients (1–21). There have been no consecutive surgical series on the basis of which to evaluate the range of such anomalies and the variability in their surgical management. In our experience, we have encountered a variety of cases in which systemic or pulmonary outflow tract obstruction was caused entirely or in part by accessory AV valvar tissue or anomalous attachments of the tensor apparatus. On several occasions, this problem necessitated modification of the planned surgical procedure, which illustrates the potential significance of such anomalies. To characterize the range of situations in which the AV valvar apparatus contributes to outflow tract obstruction and the surgical implications of such abnormalities, we reviewed our experience with this problem.

Methods

Between July 1992 and December 1997, a total of 21 patients with outflow tract obstruction due completely or in

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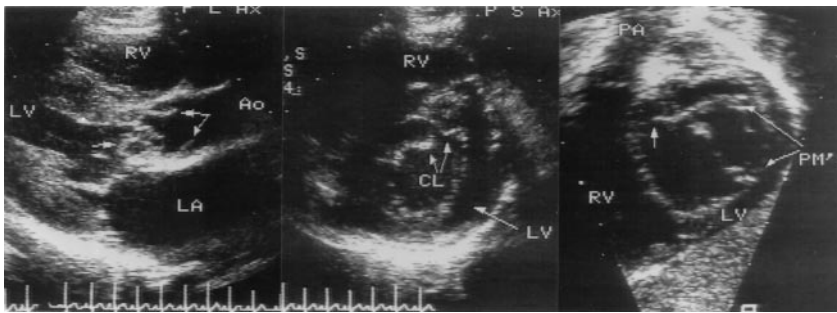


Figure 1. Preoperative echocardiogram in patient 11. **(Left)** Parasternal long axis view shows an abnormal mass of accessory AV valvar tissue (**arrow**) ballooning into the left ventricular (LV) outflow tract and across the aortic valve during systole. The open leaflets of the aortic valve are indicated by the **double arrow**. **(Middle)** Parasternal short axis view shows the abnormal septal commissure (CL) and the accessory tissue above it. **(Right)** Subcostal sagittal view demonstrates the papillary muscles (PMs) of the mitral valve, the septal commissure and the mass of accessory tissue protruding into the left ventricular outflow tract (**arrow**). Ao = aorta; LA = left atrium; PA = pulmonary artery; RV = right ventricle.

part to accessory AV valvar tissue or anomalous attachments of the tensor apparatus were evaluated for surgery at our institution. Patient data are summarized in Table 1. Ages ranged from 3 days to 19 years (median 4 years). Primary diagnoses were complex isolated systemic outflow tract obstruction ($n = 5$), congenitally corrected transposition ($n = 4$), previously repaired AV septal defect ($n = 3$), functionally single ventricle heart disease ($n = 3$; right dominant unbalanced AV septal defect in two cases and tricuspid atresia in one), transposition of the great arteries with a prior Senning procedure ($n = 2$), ventricular septal defect ($n = 2$), Shone's complex ($n = 1$) and interrupted aortic arch ($n = 1$). Ten patients had undergone prior interventions, including AV septal defect repair ($n = 3$), Senning procedure ($n = 2$), palliation or repair of arch obstruction ($n = 2$), univentricular palliation with a systemic-pulmonary arterial shunt and/or bidirectional cavopulmonary anastomosis ($n = 2$) and balloon aortic valvuloplasty ($n = 1$). All patients were evaluated with preoperative echocardiography, and 12 patients underwent preoperative catheterization. Operative techniques were based on operative findings and will be discussed in the Results section.

Perioperative data were collected on retrospective review of patient records. Follow-up was obtained by contacting the referring cardiologist. Data are presented throughout as median and range.

Results

Echocardiographic and surgical findings. All patients were diagnosed by preoperative echocardiography with obstruction to the systemic outflow tract ($n = 13$), the pulmonary outflow tract ($n = 5$) or both ($n = 3$). The median outflow tract gradient was 58 mm Hg (range 20 mm Hg to 110 mm Hg). In 13 cases the left AV valve contributed to outflow tract obstruction (Fig. 1), while the right AV valve contributed in 4 cases, both AV valves contributed in 3 and a common AV valve contributed in 1 patient with a functional single ventricle. One or more additional mechanisms of outflow tract obstruction were present in 18 patients, 17 of whom had at least one additional cause of subvalvar obstruction. There was associ-

ated aortic regurgitation in three patients (moderate = 2; mild = 1) and mitral regurgitation in two (both mild). Diagnostic variables are listed in Table 1.

Operative management. Accessory tissue and anomalous septal attachments associated with the systemic AV valve were inspected through the semilunar (usually aortic) valve. In cases of accessory tissue, the valve was often inspected through a left atriotomy as well in order to ensure that resection would not compromise AV valvar function. The tissue was then resected and additional procedures were performed as necessary (Table 2). In the other patients, various procedures and approaches were used. In 13 patients, additional outflow tract myectomy was performed, and in 11 an additional component of membranous obstruction was resected. Patients underwent surgery on moderately hypothermic cardiopulmonary bypass, with aortic and bicaval cannulation. Myocardial protection was performed with cold crystalloid cardioplegia and the aorta was cross-clamped. Circulatory arrest was not employed. Transesophageal echocardiography was used for intraoperative monitoring in all patients.

Outcomes. Support was withdrawn in the patient with a severely unbalanced AV septal defect in whom univentricular palliation was refused and a Ross-Konno operation was not possible owing to systemic outflow tract obstruction by functional left AV valvar tissue, and the patient subsequently died. Postmortem examination was not obtained. Of the 20 patients who underwent surgery, there were two early deaths. One of these was a patient with right isomerism, asplenia and subvalvar obstruction of both outflow tracts who underwent a modified Damus procedure and single ventricle palliation, and suffered an unexplained cardiopulmonary arrest 2 days after surgery from which he could not be resuscitated even with a ventricular assist device. The other early death was a patient with corrected transposition and multiple bilateral chordal attachments to the rim of the inlet ventricular septal defect who underwent bidirectional cavopulmonary anastomosis. Four days after surgery he became hypotensive and bradycardic, and the combination of decreased systemic arterial pressure and elevated systemic venous pressure in the superior caval system led to decreased transcapillary pressure in the

Table 1. Demographic and Diagnostic Data for 21 Patients With Accessory of Anomalous AV Valvar Apparatus Causing Outflow Tract Obstruction

Pt	Age (years)	Diagnosis	Outflow Tract	AV Valve	Gradient (mm Hg)	Mechanism of AV Valvar Apparatus Causing Outflow Tract Obstruction	Functional AV Valvar Tissue	Other Mechanisms of Obstruction
1	10	TGA, prior Senning, subPS	P	L	100	Multiple adhesions of the anterior mitral leaflet septal and free wall aspects of LVOT	No	Membranous, dynamic septal bowing into LVOT
2	11	TGA, prior Senning, subPS	P	L	90	Accessory MV tissue	No	Dynamic septal bowing into LVOT
3	7	Interrupted arch prior palliation, subAS	S	L	55	Tricuspid pouch extruding into LVOT	No	Membranous
4	10	Corrected TGA, VSD, valvar PS, subPS, supra-valvar PS	P	L	86	Accessory MV tissue	No	Membranous, muscular, valvar
5	15	Congenital AS, subAS, aortic regurgitation, two prior balloon dilatations	S	L	20	Accessory MV tissue	No	Membranous
6	4 m	VSD, double chamber right ventricle, subPS	P	R	60	Large functional papillary muscle supplying entire anterior leaflet of TV anchored at os infundibulum, crossing RVOT	Yes	Fibrotic muscular os infundibulum, superiorly displaced
7	7 m	Right isomerism, right dominant unbalanced AVSD (single ventricle), subAS, subPS	B	C	50 subAS 36 subPS	Functional and accessory AV valvar apparatus causing subAS and subPS	Both	None
8	3	SubAS, aortic regurgitation, "cleft" MV	S	L	110	Abnormal nonfunctional MV chordae running from base of MV to septum	No	Membranous, muscular
9	9	Complete AVSD, prior repair, subAS	S	L	90	Accessory MV tissue	No	Membrane, tunnel-type muscular
10	6	Corrected TGA, apicocaval juxtaposition, VSD, valvar PS, subPS	B	B	30	1) Accessory MV tissue & functional chordae on VSD rim causing subPS; 2) MV & TV chordal attachments to rim of VSD prevented Rastelli baffle	Yes	Muscular
11	3	SubAS, aortic regurgitation	S	L	60	Accessory MV tissue attached to chordal structures of AML, at site of abnormal commissure	No	Membranous, muscular
12	3	Corrected TGA, inlet VSD, valvar PS, subPS	S	B	100	Multiple functional MV & TV chordae crossing & attaching to rim of inlet VSD prevented Rastelli baffle	Yes	Valvar
13	7 d	Coarctation, subAS, "cleft" MV	S	L	30	Accessory MV tissue	No	None
14	4	Corrected TGA, VSD, valvar PS, subPS	B	B	45	1) Accessory MV tissue causing subPS; 2) TV chordal attachments to rim of VSD prevented Rastelli baffle	Both	Muscular, valvar
15	3 d	Right dominant unbalanced AVSD (borderline single ventricle), valvar AS, subAS	S	L	no antegrade flow	Accessory & functional MV tissue causing subAS (only retrograde flow in ascending aorta), precluded Ross-Konno	Both	Membranous, hypoplastic LV (muscular), valvar
16	14 m	Complete AVSD (non-Downs), azygos continuation of IVC, prior repair, subAS	S	L	80	Abnormal nonfunctional MV chordal structure & papillary muscle running from base of MV to septum	No	Membrane, tunnel-type muscular
17	6	Complete AVSD, double-orifice left AV valve, prior repair, subAS	S	L	45	Redundant nonfunctional chordae attaching to abnormal papillary muscle and prolapsing into LVOT	No	Membrane, tunnel-type muscular
18	3	SubAS, aortic regurgitation	S	L	55	Abnormal nonfunctional MV chordal structure & papillary muscle running from base of MV to septum	No	Membranous, muscular
19	6 d	Coarctation, subAS	S	R		TV chordal attachments to infundibular septum (interfered with subAS repair)	Yes	Muscular
20	4	Tricuspid atresia, d-transposition, subAS, prior single ventricle palliation	S	R	80	Residual TV tissue	No	Restrictive VSD
21	19	VSD, subPS	P	R	35	Tricuspid pouch extruding into RVOT during systole	No	None

AS = aortic stenosis; AV(SD) = atrioventricular (septal defect); C = common; L = left; LVOT = left ventricular outflow tract; MV = mitral valve; P = pulmonary; PS = pulmonary stenosis; R = right; RVOT = right ventricular outflow tract; S = systemic; TGA = transposition of the great arteries; TV = tricuspid valve; VSD = ventricular septal defect.

Table 2. Operative and Postoperative Data in Patients Undergoing Repair of Outflow Tract Obstruction Due to AV Valvar Anomalies

Pt	Surgical Management of Accessory/Anomalous AV Valvar Tissue	Other Procedures	Residual Gradient	Follow-up Duration (months)
1	Division of abnormal chordae and reattachment to anterior MV leaflet, release of adhesions between LVOT and anterior MV leaflet	LVOT myectomy, LVOT membrane resection	30	65.7
2	Resection of nonfunctional valvar tissue	None	30	57.3
3	Resection of nonfunctional valvar tissue	LVOT membrane resection	5	57.1
4	Resection of nonfunctional valvar tissue	LVOT myectomy (extensive), LVOT membrane resection, PV commissurotomy, supravalvar resection	15	56.4
5	Resection of nonfunctional valvar tissue	Ross procedure, LVOT myectomy, LVOT membrane resection	0	42.8
6	Anomalous papillary muscle preserved, muscle resected from around base of papillary muscle	Circumferential RVOT myectomy, infundibular patch, VSD closure	5	40.9
7	DKS, resection accessory AV valve tissue causing subpulmonic obstruction	Subpulmonary myectomy	Converted to Damus	Early death
8	Division of nonfunctional chordae	LVOT myectomy (extensive), LVOT membrane resection	0	29.7
9	Resection of nonfunctional valvar tissue	LVOT myectomy (extensive), LVOT membrane resection, AV plasty (commissurotomy, thinning)	10	27.5
10	Double switch not performed; VSD closed with RV-Ao & LV-PA continuity	None	20	26.1
11	Resection of nonfunctional valvar tissue	LVOT myectomy (mild), LVOT membrane resection	0	25.8
12	Double switch not performed; BCPA (instead of LV-PA conduit), leaving option of double switch with mechanical valves in future	Bidirectional cavopulmonary anastomosis, PA ligated and divided, atrial septectomy	PA ligated	Early death
13	Resection of nonfunctional valvar tissue	Closure of septal MV commissure ("cleft")	0	Late death (6 months)
14	Double switch not performed; VSD closed with RV-Ao continuity, LV-PA conduit placed to circumvent subPS	PA ligated and divided, VSD closed, LV-PA conduit placed	PA ligated	20.6
15	Parents refused Norwood operation, support withdrawn	n.a.	n.a.	n.a.
16	Resection of nonfunctional valvar tissue	LVOT myectomy (extensive), LVOT membrane resection	20	15.1
17	Resection of nonfunctional chordae and papillary muscle	LVOT myectomy (extensive), LVOT membrane resection	8	12.4
18	Resection of nonfunctional valvar tissue	LVOT myectomy (extensive), LVOT membrane resection	0	11.3
19	SubAS resection, not completely possible due to TV chordal attachments; Ross-Konno	LVOT myectomy (inadequate), Ross-Konno	0	9.2
20	Resection of nonfunctional valvar tissue	VSD enlargement	0	7.1
21	Resection of nonfunctional valvar tissue	VSD closure	0	1.0

Ao = aorta; AV = aortic valve; BCPA = bidirectional cavopulmonary anastomosis; LV = left ventricle; PA = pulmonary artery; RV = right ventricle. Other abbreviations as in Table 1.

cerebral circulation and brain damage. He subsequently developed multiorgan failure and support was eventually withdrawn.

In 14 patients, the accessory tissue or anomalous attachments of the AV valvar apparatus were resected completely or divided, respectively. In 10 of these patients, the postoperative outflow tract gradient was ≤ 10 mm Hg. In two patients, there was residual muscular outflow tract obstruction with gradients of 15 mm Hg and 20 mm Hg. In the two patients with transposition and a prior Senning procedure, there was a residual gradient of 30 mm Hg due to systolic bowing of the interventricular septum into the left ventricular outflow tract.

In one patient with right isomerism and a functionally single ventricle heart, only partial relief of obstruction was obtained. There was obstruction of both outflow tracts by functional chordae and accessory tissue, and despite a modified Damus procedure, this patient did not survive.

In five patients, the planned surgical procedure could not be performed because of anomalous attachments of functional

AV valvar apparatus interfering with outflow tract reconstruction. Three of these patients had corrected transposition of the great arteries, and the plan was to attempt a double-switch procedure if possible, rerouting venous return with a Senning or Mustard baffle and ventricular outflow by means of either an arterial-switch procedure or a Rastelli intraventricular baffle (22). Anomalies of the pulmonary valve (valvar stenosis in two and a bicuspid valve with mild regurgitation in the other) precluded arterial switch, leaving a Rastelli procedure as the only option. However, multiple functional left and right AV valvar chordae were attached to the rim of the ventricular septal defect, effectively precluding a Rastelli procedure without simultaneous replacement of the AV valves. Because the chordal attachments were bilateral, multiple, and diffuse (that is, not into a single papillary muscle), it was not possible to translocate the chordae into the proper ventricle, as we have done with straddling AV valve (23), or to utilize the REV procedure/conal flap method (24,25). The risk of bilateral AV

valvar dysfunction was unacceptably high. Instead of performing a double switch, all three of these patients underwent the traditional form of repair of corrected transposition, with the right ventricle retained as the systemic ventricle and pulmonary outflow obstruction relieved with a left ventricle-pulmonary arterial conduit.

There were two other situations in which anomalous attachments of the AV valvar apparatus necessitated a modification of the surgical procedure. In one patient, a Ross-Konno procedure was required because chordal attachments to the outlet septum limited the extent of infundibular septal resection that could be achieved (26). In a patient with double-chambered right ventricle, right ventricular outflow tract obstruction was caused by a large functional papillary muscle that supplied the entire anterior tricuspid valvar leaflet and inserted into the os infundibulum. A significant amount of infundibular muscle was resected, including around the base of the anomalous papillary muscle, but a substantial gradient remained after discontinuation of bypass and evaluation by transesophageal echocardiography. The patient was placed back on bypass, additional muscle was resected and an infundibular patch was placed to augment the outflow tract, resulting in complete relief of obstruction.

Follow-up. Patients were followed for 1 to 66 months (median 27 months) postoperatively. There was one late death, 6 months after surgery, in a patient with Shone's complex who required two subsequent operations for supra-aortic stenosis (5 months later) and mitral stenosis and regurgitation (6 months later). No other patients underwent reoperation during the follow-up period. On follow-up echocardiographic evaluation, the Doppler-estimated pressure gradient across the affected outflow tract (Table 2) was unchanged in all surviving patients, and no patient had evidence of new or increased AV valvar dysfunction compared with the preoperative period.

Discussion

We have presented here our experience with 21 patients in whom accessory AV valvar tissue or anomalous attachments of the AV valvar apparatus contributed to outflow tract obstruction or interfered with outflow tract reconstruction. In the majority of cases, the outflow tract of the morphological left ventricle was involved, which reflects the critical role of the left AV valve in the left ventricular outflow tract. Although accessory or anomalous AV valvar tissue was the predominant cause of outflow tract obstruction in most patients, the obstruction was typically complex, with multiple contributing factors, including muscular ($n = 14$), membranous ($n = 11$) and dynamic ($n = 2$) obstruction as well. Consistent with most previously reported cases (1-21), we found that obstruction due to nonfunctional (accessory) tissue can almost always be relieved completely as long as it is identified. In contrast, if functional tissue interferes with outflow tract repair, it has much more significant implications for the approach to repair and possibly for outcome.

Accessory/anomalous AV valvar tissue causing outflow tract obstruction. *Accessory AV valvar tissue.* In the majority of our cases, accessory tissue or anomalous attachments of the AV valves contributed to actual outflow tract obstruction, as opposed to interfering with outflow tract reconstruction. In hearts with normal topology and segmental connections, the most common finding was accessory (nonfunctional) left AV valvar tissue contributing to subaortic obstruction. In two patients, this tissue took the form of a sail-like membrane with chordal attachments to the papillary muscles that would fill with blood during systole and balloon into the left ventricular outflow tract. This type of accessory mitral valve tissue has been described in several reports (1,3,11), and is important because it can cause severe outflow tract obstruction in the dynamic heart, but assumes a less obvious profile in the intraoperative state of chamber relaxation and decompression. In the pre-echocardiography era, there were instances in which such tissue was overlooked on operative inspection, with a fatal consequence (1). With echocardiography, this pattern can be recognized quite readily (Fig. 1). Other than this pattern, accessory tissue in our experience and in the literature seems to constitute a spectrum, which typically has the appearance of mature AV valvar tissue on histopathologic examination (11). In our patients, accessory AV valvar tissue was typically associated with some degree of membranous and/or muscular subaortic obstruction as well.

Accessory tissue of the left AV valve appears to be a relatively common cause of left ventricular outflow tract obstruction in both transposition and AV septal defect as well (8,17,18), and should not pose any different management problems than in cases of isolated subaortic obstruction. As in the various case reports describing this phenomenon, resection of the accessory tissue, along with resection of membranous or muscular substrata of obstruction, allowed for essentially complete relief of outflow tract obstruction in all of our cases, with no evidence of resultant AV valvar dysfunction (3,6,7,10,11). We are not aware of any reported complications of resecting accessory AV valvar tissue, although failure to identify such tissue has been known to lead to persistent outflow obstruction and death (1,5-7,19).

In two patients with normally related great arteries, systemic or pulmonary outflow tract obstruction was caused by a tricuspid pouch extruding into either the left or right ventricular outflow tract, respectively, during systole. The patient with subaortic obstruction according to this mechanism had elevated right ventricular pressures after prior palliation for interrupted aortic arch at another institution with pulmonary arterial banding and a pulmonary trunk to descending aortic conduit. Outflow tract obstruction due to a tricuspid pouch (sometimes referred to erroneously as a ventricular septal aneurysm) has been described in patients with both normally related and transposed great arteries (8,9,15,16). Rare cases of actual ventricular septal aneurysm causing outflow tract obstruction have also been reported (4,20). Closure of the ventricular septal defect, along with resection of any accessory tissue, sufficiently relieves obstruction due to this mechanism.

Anomalous attachments of the tensor apparatus. Anomalous attachments of the subvalvar AV valvar apparatus causing outflow tract obstruction have been described in a number of reports, both as an isolated anomaly and in association with other types of congenital heart disease (1,2,12). The most common circumstance appears to be AV septal defect or isolated "cleft" mitral valve, in which chordae run from the edges of the septal commissure ("cleft") to the septum. These can be divided, as in two patients in our series, and the septal commissure closed without causing AV valvar regurgitation. Attachments of abnormal chordae to the septal wall of the left ventricle have been described in patients without AV septal defect or "cleft" mitral valve as well, including two in this series, one with transposition and another with isolated left ventricular outflow tract obstruction. As long as the valve is sufficiently supported by normal tensor apparatus, these anomalous attachments can be divided and resected. However, several reports have described an abnormal papillary muscle and chordae inserting into the base of the anterior mitral valvar leaflet with no normal chordae from the free edge of the leaflet (13). In such cases, resection of the abnormal component of the subvalvar apparatus and replacement of the valve or insertion of artificial chordae (expanded polytetrafluoroethylene suture) are essentially the only options.

Anomalous attachments of the left AV valve to the septum may be an important cause of subpulmonary outflow tract obstruction in complete transposition of the great arteries as well. Among 166 specimens in the Edwards collection, 21 had subpulmonary (left ventricular) outflow obstruction, 7 of which had anomalous attachments of the left AV valve to the septum (2). In patients with a Senning procedure and leftward septal bowing, the combination of these effects may lead to severe obstruction, as in one of our patients.

Anomalous AV valvar tissue interfering with outflow tract reconstruction. Several types of anomalous AV valvar tissue interfered with outflow tract reconstruction. In all but one case, we were forced to abandon our planned outflow tract reconstruction in favor of an approach that would not be compromised by the AV valvar anomalies. In the other case, modification of the planned repair was required.

Attachments of functional tensor apparatus to the crest of the septum in transposition complexes. The most common abnormality in this category was the presence of multiple left and right AV valvar chordae diffusely attaching to or crossing the rim of the ventricular septal defect. In three patients with corrected transposition, this precluded a Rastelli intraventricular baffle procedure as part of a double-switch operation (22). These patients all had abnormalities of the pulmonary valve, similarly precluding arterial switch. As a result, they were managed either with standard repair of corrected transposition, leaving the right ventricle as the systemic ventricle, or with bidirectional cavopulmonary anastomosis in order to preserve the option of anatomic repair with a mechanical AV valve at a later date. The fourth patient with corrected transposition in this study had severe valvar pulmonary steno-

sis and a small ventricular septal defect, so a double-switch procedure was not planned.

Chordal attachments to the crest of the ventricular septal defect are not uncommon in patients with transposition complexes such as corrected transposition, complete transposition, and double-outlet right ventricle (24,25). If the pulmonary valve is normal and arterial switch is an option, the anomalous chordal attachments should not have significant implications for outflow tract reconstruction if these attachments are not significantly obstructive. If the valve is not normal, however, such anomalous chordal attachments can be critical. An option that has been employed to baffle the left ventricle to the aorta in patients with transposition or double-outlet right ventricle, pulmonary stenosis and discreet chordal bundles inserting near or on the rim of the ventricular septal defect is the REV procedure, or conal flap method. With this technique, the anomalous chordal insertions are raised from the septum as one or two pedicled flaps of septal wall based away from the defect, and the outlet septum is resected, after which the Rastelli baffle is performed and the flap(s) are secured to the right ventricular aspect of the baffle patch (24,25). In properly selected patients, this method appears to be effective, with a small but important risk of tricuspid regurgitation. However, it is not an option in patients such as ours, in whom the chordal attachments were multiple, diffuse and bilateral.

A technique similar to the conal flap method, which we have employed for biventricular repair in patients with AV valvar straddling, is translocation and realignment of anomalously located chordal bundles (23). This approach may also be useful in cases of discrete chordal bundles inserting into the septum at the rim of the defect, but cannot be used safely with the type of complex attachments seen in these cases. Aside from abandoning the Rastelli procedure, the only option for overcoming the anomalous AV valvar insertions in cases with diffuse, multiple and/or bilateral chordae attaching to or crossing the rim of the defect appears to be valve replacement, which is a suboptimal choice in children.

Other forms of functional AV valvar apparatus interfering without outflow tract repair. In four other cases, the AV valvar tensor apparatus interfered with potential outflow tract reconstruction. A Ross-Konno procedure was either necessary because chordal attachments to the outlet septum limited the extent of infundibular septal resection that could be achieved ($n = 1$), or was not possible owing to functional AV valvar apparatus obstructing the subaortic region ($n = 1$). The Ross-Konno procedure has proved to be a very important addition to the repertoire of the cardiac surgeon managing systemic outflow tract disease (26). As these two cases illustrate, the Ross or Ross-Konno operations may increase the options for treatment of outflow tract obstruction due to anomalies of the AV valvar apparatus, but such anomalies may also interfere with planned reconstruction using the Ross-Konno procedure.

In a patient with right isomerism, multiple chordae from the common AV valve were obstructing both outflow tracts, and the patient died after a modified Damus-Kaye-Stansel proce-

ture, with residual outflow obstruction. Anomalous AV valvar structures have been shown to contribute to outflow obstruction in a considerable proportion of patients with isomerism (20), although such involvement is more common in left than in right isomerism, and is often due to accessory rather than functional tissue. Unlike anomalies amenable to biventricular repair, there are few good options for managing outflow obstruction attributable to functional AV valvar apparatus in the setting of univentricular heart disease. Residual outflow obstruction and AV valvar regurgitation (a potential consequence of resecting AV valvar tissue) are not well-tolerated and can have significant implications for ventricular function, and relocation of the systemic outflow tract with a conduit (as in the Rastelli procedure) is not feasible in univentricular palliation.

Types of AV valvar apparatus causing outflow tract obstruction not included in this study. Two of the most important and most common forms of subaortic obstruction caused in part by the AV valvar apparatus have not been included in this study. Systolic anterior motion of the mitral valve in hypertrophic obstructive cardiomyopathy was not included primarily because this dynamic process has traditionally been considered a functional abnormality, ascribed to the Venturi effect in the setting of severe septal muscular obstruction, and not a structural abnormality of anomalous AV valvar tissue or attachments. It has been proposed, however, that a critical feature of subaortic obstruction due to systolic anterior motion of the mitral valve is anterior displacement of the papillary muscles (27). It is also known that elongation of the mitral valve leaflets are present in patients with this lesion. Thus, this is still considered more a functional than a structural anomaly of the AV valvar apparatus.

The other recognized form of AV valve-related outflow tract obstruction that was not included in this report was subaortic obstruction due to the narrowed and elongated left ventricular outflow tract in AV septal defect, which is in part a function of the deficient interventricular septum and consequent displacement of the base of the anterior left AV valvar leaflet (28). Although this can be managed in some cases with reconstruction of the anterior leaflet at its basal attachment (18), this problem is more a function of abnormal development of the AV septum than of the valves or valvar apparatus per se. We did include three patients with previously repaired complete AV septal defect in this series, but all three patients had abnormalities of the subvalvar apparatus that contributed to subaortic obstruction.

Although replaced or reconstructed AV valves are not native anomalies, it is worth noting that they can cause left ventricular outflow tract obstruction as well (29,30).

Conclusions. Although rare, anomalous functional and accessory AV valvar apparatus are potentially important causes of both pulmonary and systemic outflow tract obstruction. Most often, accessory tissue is involved, and is thus amenable to resection. As others have suggested (11), because accessory AV valvar tissue causing outflow tract obstruction is not a commonly observed condition and may be difficult to identify

intraoperatively in the arrested heart, it may be overlooked. Accordingly, its prevalence may be underestimated and it may contribute to residual obstruction and even death (1,5-7,19). This mechanism of obstruction is becoming recognized in an increasing number of cases, in part due to the widespread use and increasing facility with echocardiography, and should be considered regularly as a potential cause of outflow tract obstruction.

The relationship between outflow tract obstruction due to AV valvar anomalies and fibrous or muscular causes is not known. It has been proposed that turbulent flow caused by AV valvar tissue in the outflow tract may promote fibrous proliferation and membranous subvalvar obstruction. By the same token, flow disturbances and hemodynamic forces across the AV valvar tissue may stimulate growth of the accessory AV valvar tissue itself. Regardless, there are no reports of which we are aware that accessory tissue recurs after resection. Because accessory tissue can be resected completely and does not appear to redevelop, the outlook for this type of outflow tract obstruction is favorable. However, because accessory tissue is often found with discrete fibromuscular or tunnel type subaortic obstruction, patients may be subject to recurrence of these components. Conversely, patients with outflow tract obstruction due to functional AV valvar tissue may be compromised as a result, either because the obstruction cannot be relieved completely or because the repair of choice cannot be performed.

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