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Development of a Double-Chambered Right Ventricle After Repair of Tetralogy of Fallot

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Objectives. **We sought to determine the frequency, etiology and progressive nature of midcavity obstruction in patients after primary repair of tetralogy of Fallot (TOF).**

Background. **Midcavity obstruction (double-chambered right ventricle [DCRV]) represents a significant portion of reoperations in patients who have had TOF repair. This group is still poorly defined.**

Methods. **A retrospective review of clinical, echocardiographic and catheterization data for all patients with TOF who later underwent reoperation for DCRV was performed.**

Results. **Between 1973 and 1995, 552 children <2 years of age underwent primary TOF repair (median age 6.7 months). Long-term follow-up (median 50 months) was available in 308 children. Of these, 17 children subsequently developed DCRV requiring reoperation. The median age at initial operation was 7.9 months. During a median follow-up interval of 43.2 months, murmur intensity increased in all patients, and the average subpulmonary gradient at catheterization increased from 24 ± 10 to 80 ± 27 mm Hg in seven children (p = 0.002) and at Doppler echocardiography from 14 ± 16 to 89 ± 18 mm Hg in five children (p = 0.002). Before reoperation, 6 of the 17 children were symptomatic. During the operation (median age 55.4 months), obstruction was relieved by incision of hypertrophied anomalous muscle bundles in all 17 patients, with prominent fibrosis noted in 8 patients. Excessive septal and parietal hypertrophy was noted in one child. No new transannular patches were required. Recurrent obstruction has reappeared in 3 of these 17 children during follow-up.**

Conclusions. **DCRV is a medium-term complication of TOF repair in infants, with a minimal incidence of 3.1% (95% CI 1.8% to 4.9%). The condition is progressive and is due to anomalous muscle bundle hypertrophy or fibrosis, or both, which may represent displaced insertion of a moderator band. Further reobstruction does occur; continued careful follow-up is therefore essential.**

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Tetralogy of Fallot (TOF) is the most common cyanotic heart lesion, occurring in 8% to 9.7% of children with congenital heart disease (1,2). Over the past two decades complete repair of this lesion, even in neonates, has become routine in many centers. As part of this approach, limited ventriculotomy and outflow muscle resection with patch augmentation are employed (3). Using this technique, the need for reoperation, including subsequent subpulmonary/midcavity obstruction requiring surgical relief, has occurred in 0.05% to 9% of patients (4–6).

The purpose of this retrospective study was to determine the minimal frequency of subpulmonary/midcavity obstruction among patients with TOF repaired in the first 2 years of life during the past two decades at our institution. Because TOF, in itself, is associated with subpulmonary stenosis secondary to a displaced conal septum, we sought 1) to better define the etiology of subpulmonary obstruction in those patients with restenosis; 2) to distinguish between recurrent infundibular obstruction and double-chambered right ventricle (DCRV) (midcavity obstruction due to a displaced moderator band) and; 3) to determine whether such obstruction was progressive.

Methods

Patients. The computerized data base was first searched for all patients with the diagnosis of TOF who underwent primary repair in the first 2 years of life from 1973 to 1995. This group was then investigated for those who underwent subsequent reoperation for subpulmonary/right ventricular outflow obstruction at our institution. All available clinical, electrocardiographic, echocardiographic, cardiac catheterization and surgical data were reviewed in detail and form the basis of this report. In addition, the typical position of the moderator band was defined in 20 consecutive patients <2 years of age who underwent repair of TOF without developing DCRV, and an attempt was made to use the previously reported moderator to pulmonary valve distance/tricuspid valve annulus ratio, which

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Abbreviations and Acronyms

CI = confidence interval
DCRV = double-chambered right ventricle
TOF = tetralogy of Fallot
VSD = ventricular septal defect

distinguishes patients with a DCRV/ventricular septal defect (VSD) from those with an isolated VSD or normal patients (7).

Statistical data. Age and follow-up intervals are expressed as mean value ± SD and median. Proportions are expressed with 95% confidence intervals (CI). The Student *t* test and extended Fisher exact test were applied where appropriate. A p value <0.05 was considered significant.

Results

Of the 552 patients who underwent primary repair of TOF in the first 2 years of life, follow-up (median 50 months) was available in 308 through our data base. Of these, 48 (5.5%, 95% CI 3.2% to 8.7%) underwent reoperation at our institution, including 17 (35%) with DCRV (7 male and 10 female), who are the subject of this report. Other reoperations included three pulmonary valve replacements, three right ventricle to pulmonary artery conduits, five main pulmonary artery plasties, eight right pulmonary artery plasties, nine left pulmonary artery plasties, seven VSD revisions, four aneurysmectomies, four patent foramen ovale closures, five wire removals and one correction of partial anomalous venous return.

Table 1. Data at First Operation

Pt No./Age (mo)	Echo	Cath	PA Anat	Patch Type	Arch Type
1/14.9	—	M	N	NTA	LAA
2/2.7	—	T	N	TA	LAA
3/7.1	—	M	N	NTA	LAA
4/11.1	MB	M	RP/MP	NTA	LAA
5/4.5	MB	T	N	NTA	RAA
6/10.0	—	M	N	NTA	RAA
7/11.6	—	T	RP/MP	TA	LAA
8/7.9	—	M	N	NTA	LAA
9/7.9	—	M	N	NTA	LAA
10/15.6	—	T	N	TA	LAA
11/17.4	—	T	N	NTA	LAA
12/16.3	—	M	N	NTA	LAA
13/3.7	MB	T	N	NTA	LAA
14/2.8	MB	—	—	TA	RAA
15/6.8	—	T	N	TA	LAA
16/6.8	—	T	N	TA	LAA
17/5.7	MB	T	N	TA	RAA

Cath = catheterization; Echo = echocardiography; LAA = left aortic arch; M = mild form of TOF (see text); MB = muscle bundle seen at os infundibulum; N = good-sized pulmonary arteries (PAs); NTA = nontransannular patch; Pt = patient; RAA = right aortic arch; RP/MP = main pulmonary artery angled to the right pulmonary artery, unlike normal main pulmonary artery to left pulmonary artery orientation; T = typical form of TOF (see text); TA = transannular patch.

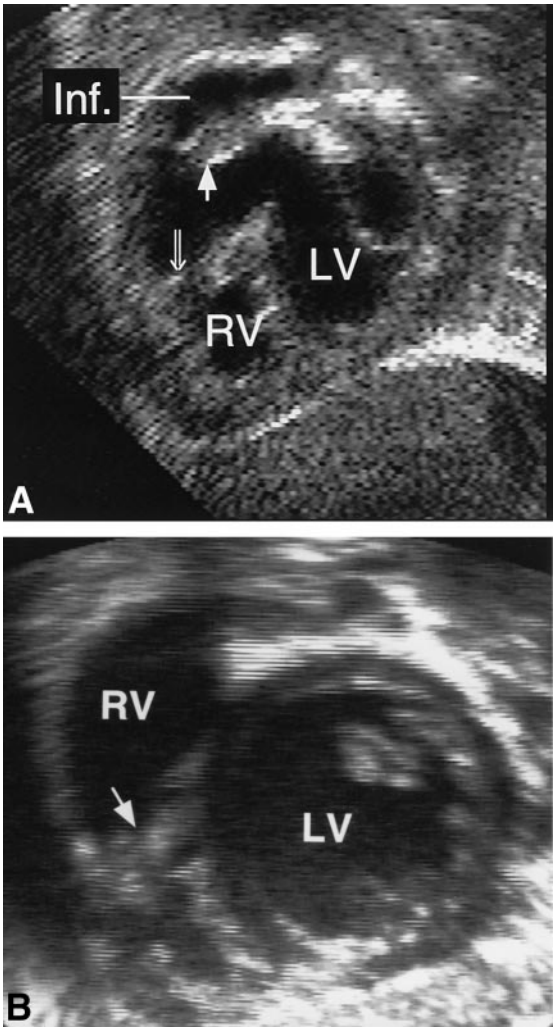

Figure 1. The subxyphoid short-axis view from a patient with TOF revealed (**A**) a displaced moderator band (**open arrow**) with more rightward and superior insertion into the interventricular septum; anterior displacement of the conal septum is also shown (**solid arrow**). This has a similar appearance to the typical position of the moderator band in DCRV associated with a membranous VSD (**B**). Inf. = infundibulum; LV = left ventricle; RV = right ventricle.

Data before and at initial repair (Table 1). Echocardiographically, because of the era involved, only five of the 17 preoperative TOF studies were available for review. All revealed features consistent with the diagnosis of TOF and, in addition, had evidence of a displaced moderator band rightward and superiorly to the anteroinferior ridge of the VSD (as seen in DCRV) (Fig. 1). Doppler interrogation at the time revealed no significant intracavitary obstruction. Cardiac catheterizations, undertaken in 16 of the 17 patients, revealed two distinct morphologic findings angiographically: 1) a typical anterior malaligned VSD (n = 9) with a small infundibular chamber and a short, small main pulmonary artery in an almost horizontal plane on the lateral projection (8); and 2) a milder form of TOF (n = 7) with a larger, more superiorly oriented main pulmonary artery. The branch pulmonary arteries were of good size in all patients, with the main pulmonary artery

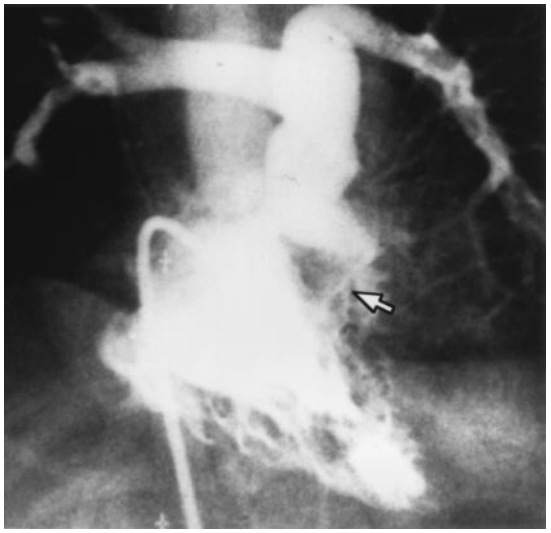

Figure 2. The anteroposterior projection of the right ventricle in a patient with TOF illustrates the displaced moderator band **(arrow)** that later became hypertrophied after primary repair.

angled toward the right in two patients. Prominent anomalous muscle bundles, best seen in the anteroposterior view (Fig. 2), were evident in 71% of patients, and conal hypertrophy in an additional 21%.

The age and weight of these 17 patients at primary repair were 9.0 ± 4.8 months (median 7.9) and 7.3 ± 1.5 kg (range 1st to 75th percentile). The outflow patch placed was nontransannular in 10 patients and transannular in 7. During this operation, anomalous muscle bundles requiring transection/resection were described in six patients, excessive trabeculation in two and high papillary muscle insertion in one. Pulmonary artery pullback on the first postoperative day revealed a gradient of 8.4 ± 5.7 mm Hg (median 8.5) in 10 patients.

Interval data (Table 2). Reobstruction was initially suspected because of an increase in the systolic ejection murmur (grade ≥4 in 15 patients). Electrocardiograms, available in 14 patients, indicated right ventricular hypertrophy in 10. Serial catheterizations in seven patients revealed progressive obstruction in all, with a peak systolic ejection gradient of 24 ± 10 mm Hg at the initial postoperative catheterization, increasing to 80 ± 27 mm Hg ($p = 0.002$) before reoperation. Residual VSDs were noted in two patients without significant left to right shunting. In addition, five patients had serial echocardiograms that confirmed the progressive nature of this subpulmonary obstruction, revealing an increase in the maximal instantaneous gradient (14 ± 16 to 89 ± 18 mm Hg, $p = 0.002$).

Reoperation data. Mean patient age at reoperation was 68.8 ± 59.7 months (median 55.4), giving an interval-free period of 59.8 ± 58.0 months (median 43.2). Before reoperation, echocardiographic data adequate for detailed analysis were available in 12 of 17 patients. In all patients, the right ventricular outflow tract obstruction originated in the midcavity with a Doppler-derived maximal instantaneous gradient of 75 ± 18 mm Hg. In an attempt to ascertain moderator band anatomic differences, 20 consecutive patients with TOF (<2 years of age) were evaluated. In these the plane of the moderator band was more leftward and inferior (as in the normal heart) than in those who later developed DCRV.

Table 2. Follow-Up Results Between Operations

Pt. No.	Murmur	ECG RVH	Symptom	PSEG (mm Hg) at Cath	MIG (mm Hg) at Echo	PA (mm Hg) Pullback*	VSD
1	5/6	Y	Fatigue	87	72	15	N
2	3/6, harsh	Y	Cyanosis	42	—	—	Y
3	5/6	N	Asymptomatic	—	64	5	N
4	5/6	N	Asymptomatic	—	100	0	N
5	4/6	Y	Asymptomatic	80	110	—	N
6	3/6, harsh	Y	DOE	57	50	—	N
7	5/6	Y	Asymptomatic	65	60	—	Y
8	5/6	N/A	Asymptomatic	86	—	6	N
9	5/6	Y	DOE	130	80	—	N
10	5/6	Y	Asymptomatic	85	90	—	N
11	4/6	N	FTT	61	—	10	N
12	4/6	N/A	Asymptomatic	58	70	—	N
13	5/6	N/A	N/A	100	85	0	N
14	4/6	Y	Asymptomatic	82	64	12	N
15	5/6	N	Cyanosis	62	60	7	N
16	4/6	Y	Asymptomatic	60	—	15	N
17	5/6	Y	Asymptomatic	60	—	14	N

*On postoperative day 1. DOE = dyspnea on exertion; ECG = electrocardiographic; FTT = failure to thrive; MIG = maximal instantaneous gradient; N = no; N/A = not available; PSEG = peak systolic ejection gradient; Pt. = patient; RVH = standard criteria or suggested right ventricular hypertrophy by prominent R′ in lead V_1 in the presence of right bundle branch block; VSD = anatomically small ventricular septal defect without significant left to right shunt; Y = yes; other abbreviations as in Table 1.

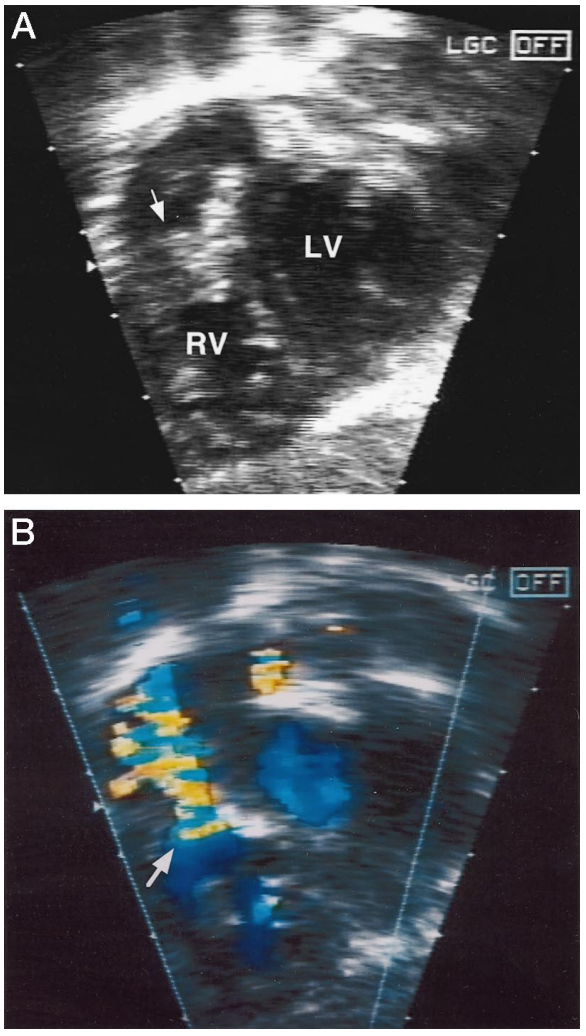

Figure 3. A, The subxyphoid view in a patient after repair of TOF. The prominent muscle bundle is shown **(arrow)** with a typical DCRV appearance. **B,** Color Doppler interrogation illustrates the midcavity obstruction. LV = left ventricle; RV = right ventricle.

However, given the retrospective nature of the study, usual TOF anatomy and apical insertion of the moderator band, it was not possible to use the ratio of normal moderator to pulmonary valve distance/tricuspid valve annulus previously described by other investigators to differentiate normal subjects from those with VSD/DCRV (7). No single frame allowed for moderator band to pulmonary valve distance measurement. In all 12 patients with DCRV/TOF, the echocardiogram showed a rightward and superiorly displaced insertion of the moderator band, giving an appearance of DCRV (Fig. 3). In three patients, an additional site of obstruction was identified—a hypertrophied conal septum or muscle in the infundibulum in one patient, severely hypertrophied trabeculae within the right ventricular outflow tract in another and thickened pulmonary valve leaflets–posttransannular patch in the third. Preoperative catheterization, undertaken in 15 patients, revealed a peak systolic ejection gradient of 74 ± 22 mm Hg and angiography identified midcavity obstruction (Fig. 4).

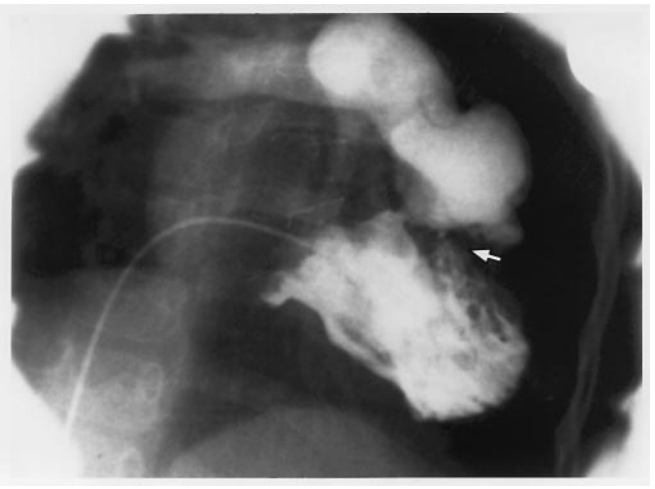

Figure 4. The anteroposterior projection after repair of TOF, with an appearance typical of DCRV with midcavity obstruction at the level of the displaced moderator band **(arrow)**.

At reoperation right ventricle cavity obstruction was due to hypertrophied anomalous muscle bundles/displaced moderator band in all patients, with prominent fibrosis noted additionally in eight. A significant contribution to the obstruction was due to septal hypertrophy in one patient. No patient required extension of a nontransannular patch to a transannular patch. No pulmonary artery stenosis was noted.

Follow-up after reoperation (Table 3). Follow-up data are available in all 17 patients, one of whom required further surgical repair consisting of a Konno procedure, including aortic valve replacement for significant left ventricular outflow tract obstruction. It is of interest that right ventricular outflow obstruction recurred in three patients (Patients 5, 8 and 13) (median follow-up 100 months), all with a murmur of increasing intensity confirmed echocardiographically in two and at catheterization in the other. In Patients 5 and 8, the obstruction appeared to be related to an increasingly prominent muscle bar. In Patient 13, a limited moderator band resection was done because of papillary muscle insertion in the conal septum; at catheterization the obstruction was both subvalvar and valvar, and with balloon dilation the PSEG decreased from 52 to 20 mm Hg.

Discussion

Anatomy. TOF and DCRV are considered by many to be two distinct lesions anatomically, the former consisting of anterior and leftward deviation of the conal septum with subvalular and valular pulmonary stenosis, an anterior malaligned VSD and aortic override (9). It is the displacement of the hypertrophied conal septum and hypoplasia of the subpulmonary region that produce the subpulmonary stenosis in TOF. In contrast, DCRV is a form of right ventricular outflow obstruction characterized by muscular narrowing that partitions the right ventricle into an inflow and outflow chamber. Wong et al. (7), in an echocardiographic and pathologic study,

Table 3. Follow-Up Data

Pt. No.	Age at Op. 1 (mo)	Age at Op. 2 (mo)	Follow-Up Interval* (mo)	Symptom	Weight (percentile)	Height (percentile)	Echo	Murmur
1	14.9	55.4	201	A	< 5	< 5	RV 25	2/6
2	2.7	58.3	181	A	25	10	RO 20	3/6
3	7.1	48.7	129	A	85	90	RO 0	3/6
4	11.1	14.4	103	A	25	10	N/A	1/6
5	4.5	47.7	100	A	40	20	RO 64	5/6†
6	10	229.1	232	A	40	70	RV 0	2/6
7	11.6	156.6	219	A	5	< 5	LPs[1]	2/6
8	7.9	63.3	220	A	85	25	RO 0	0/6
9	7.9	63.3	221	A	80	95	RV 55	4/6†
10	15.6	71.8	182	A	50	55	LPs[2]	3/6
11	17.4	49.3	190	A	80	40	RO 30	2/6
12	16.3	168.1	210	A	25	< 5	RO 0	1/6
13	3.7	10.1	55	A	75	75	CVp	5/6†
14	2.8	23.2	72	DE/m	50	5	RV 25	3/6
15	6.8	22.7	125	A	75	25	RV 20	2/6
16	6.8	31.6	207	A	75	50	RO 25	2/6
17	5.7	55.9	67.9	A	95	95	N/A	2/6

*Age (months) at last follow-up (all patients alive as of September 1996). †Recurrent obstruction. A = asymptomatic; CVp = gradient present across right ventricular outflow tract, noted on echocardiography, confirmed at catheterization, requiring valve dilation (see text for details); DE/m = dyspnea on exertion (mild); LPs[1] = LPA stenosis 16 to 20 mm Hg on echocardiogram; LPs[2] = LPA stenosis <8 mm Hg on echocardiogram; N/A = not available; Op. 1 = Age at initial tetralogy of Fallot repair; Op. 2 = Age at operation for double-chambered right ventricle; Pt. = patient; RO = right ventricular outflow tract obstruction (maximal instantaneous gradient, mm Hg); RV = right ventricular pressure by tricuspid regurgitant jet (mm Hg).

found that the obstruction in DCRV is due to a hypertrophied moderator band with superior and rightward displacement of its insertion into the septal band. DCRV is characterized by progressive midcavity right ventricular outflow obstruction (10), with the presentation extending beyond the newborn period (11–13). Although DCRV is frequently associated with a membranous VSD and occasionally subaortic stenosis (10,14), a few earlier small series have documented DCRV in the presence of an anterior malaligned VSD (15–17), similar to the patients in this report.

Reoperation in TOF. Reoperation is necessary in some patients after primary repair for TOF. Since 1973, we have performed elective primary repairs in 552 children <2 years of age. Primary follow-up was conducted in 308 patients at our institution, reflecting its quaternary referral base. In this age group, there is less septal and parietal band hypertrophy, and the need for extensive resection of muscle is reduced (3). In this report, 17 of these 308 patients over a median postoperative follow-up of 43.2 months required reoperation for DCRV-like anatomy, an incidence of 5.5% (95% CI 3.2% to 8.7%). This represented 35% of all patients requiring reoperation at our institution. The incidence of reoperation in other series ranges from 3.5% to 11% (6,18–25). This is a time-related phenomenon, with 10-, 20- and 30-year freedom from reoperation being 95% to 97%, 92% and 88%, respectively (19,21,22). Of those patients undergoing reoperation, right ventricular outflow obstruction represents 6% to 67%, residual/additional VSD repair 1.5% to 74% and pulmonary valve replacement 0% to 59%. Perhaps the reason for a higher proportion of DCRVs in our patient group reflects our philosophy regarding TOF repair and the need for preservation of right ventricular function. Recent attention has focused on the factors determining late right ventricular dysfunction (26). We believe these factors include the length of ventriculotomy, preservation of the right coronary branches, the width of the transannular patch and excessive division of muscles within the right ventricle, in particular division of the moderator band. Early complete repair with limited muscle resection seems to reduce the occurrence of late right ventricular dysfunction. Of our total 552 patients, 308 were followed primarily at our institution, 48 having undergone reoperation at our institution over this period. Of these, only six patients (1.9%) are known to have required pulmonary valve replacement or right ventricle to pulmonary artery conduit replacements in an attempt to treat late right ventricular dysfunction. This may be related to our limited resection methodology, although this approach may have contributed to the later recurrence of DCRV in 17 patients (3.1%, representing the minimal incidence). We do not believe this DCRV incidence represents a learning phenomenon because there is no statistical difference for all time periods, suggesting this is most likely an unrecognized anatomic substrate (Fig. 5).

DCRV in TOF. At reoperation, in all but one of our 17 patients with DCRV, surgical findings confirmed the echocardiographic/angiographic findings. Echocardiographically, the obstruction diagnosis was best displayed in the subxyphoid

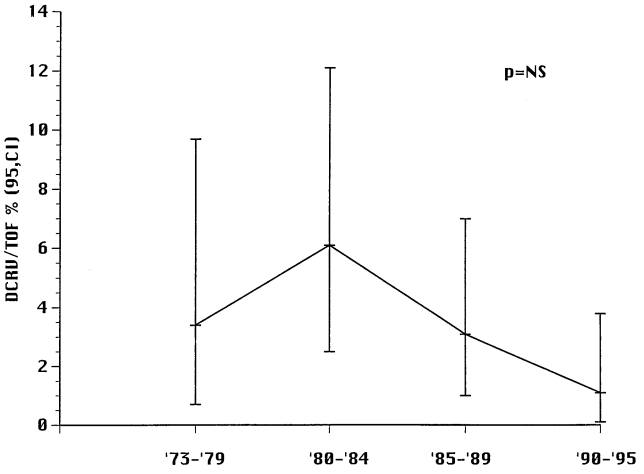

Figure 5. Patients with primary repair of TOF who later developed DCRV as a proportion of the total number of TOF operations in four time periods. Proportions and 95% CIs are shown.

short-axis view, with flow aliasing during Doppler interrogation midway into the right ventricular cavity, and angiographically it was best displayed in the frontal projection. Although the displaced moderator band may help identify the at-risk group of patients who later develop DCRV, the inciting stimulus of muscle hypertrophy remains unclear, as none of the 17 patients had more distal obstruction.

Clinical observations of increasing obstruction. Evidence of progressive obstruction was demonstrated by 1) increasing systolic murmur intensity; 2) right ventricular hypertrophy on the electrocardiogram, even in the presence of right bundle branch block in the majority of patients; and 3) gradients on the echocardiogram in five patients and at catheterization in all seven who had serial studies.

Predictors of DCRV. It is believed that extensive resection of right ventricular muscle may represent a risk factor for ventricular arrhythmias (27) and late right ventricular dysfunction (26). Early primary repair allows for minimal resection of muscle from the right ventricle (3). Because the development of DCRV may necessitate reoperation in patients undergoing early repair, a modification in our current management of at-risk patients is needed. Our study would suggest that an anatomic substrate exists, to allow for selection of the majority of patients at higher risk of subsequent DCRV development. They often have a milder form of TOF recognizable on the echocardiogram and angiogram, and the majority have evidence of an abnormally displaced moderator band septal insertion at the time of initial presentation. Echocardiographically, we were unable to apply the technique of Wong et al. (7) to compare our patients with a control group of 20 patients with TOF without DCRV, because of both a limited number of recordings in the former group and anatomic visualization constraints preventing normal TOF value determination. However, five patients with initial echocardiograms had evidence of a displaced moderator band with insertion closer to the VSD, resulting in its visualization, with the pulmonary valve in the same plane using a subxyphoid short-axis view. In addition, 12 of 16 patients with preoperative TOF angiograms had evidence of anomalous muscle bundles in the right ventricular midcavity. Subsequent preoperative DCRV echocardiograms confirmed these to be displaced moderator bands. Although operative notes in six patients describe anomalous muscle bundle resection at the time of initial repair, and recurrence of obstruction occurred in three after DCRV repair, our findings would suggest that preoperative visualization of anomalous muscle bundles does increase the risk of subsequent DCRV development. At minimum, these patients require closer follow-up. Less evidence exists for aggressive resection at the initial operation.

Study limitations. This is a descriptive report of known patients with subpulmonary obstruction after repair of TOF who presented to our institution with DCRV. The quoted incidence therefore represents the minimal incidence of this phenomenon. The number of patients requiring other forms of reoperation is again limited by the incomplete follow-up data. In addition, the frequency of displaced moderator bands in all patients with TOF and the rate of intraoperative resection rate are unknown and difficult to determine in a retrospective manner. However, to our knowledge, this is the first report presenting clinical, angiographic and echocardiographic data on the anatomic substrate that leads to DCRV in patients with TOF repair.

Conclusions. In infancy TOF repair is associated with the need for reoperation in at least 3.1% (95% CI 1.8% to 4.9%), secondary to the development of DCRV, representing up to 35% of all reoperations for TOF. The etiology of this appears to be displacement of the moderator band with subsequent progressive hypertrophy. Patients at increased risk may be identified both echocardiographically and angiographically. Because excessive resection of muscle bundles seems to be a risk factor for late right ventricular dysfunction, it seems prudent to try to identify those patients at risk for late midcavity obstruction. Within this TOF subgroup, it may be appropriate to undertake a more aggressive follow-up of these patients, even after surgical resection of muscle bundles, because this obstruction can recur. However, because of the unknown incidence of a displaced moderator band in TOF and the unknown effect of moderator band resection on right ventricular function, the clinical management of potential DCRV in patients remains unresolved and requires further investigation.

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