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Radiofrequency Catheter Ablation of Supraventricular Tachycardia Substrates After Mustard and Senning Operations for *d*-Transposition of the Great Arteries

Ronald J. Kanter, MD, FACC,* John Papagiannis, MD,§ Michael P. Carboni, MD,*
Ross M. Ungerleider, MD, FACC,† William E. Sanders, MD, FACC,‖ J. Marcus Wharton, MD, FACC‡
Durham and Chapel Hill, North Carolina, and Athens, Greece

OBJECTIVES The purpose of this study was to determine the efficacy and risks of radiofrequency ablation of various forms of supraventricular tachycardia after Mustard and Senning operations for *d*-transposition of the great arteries.

BACKGROUND In this patient group, the reported success rate of catheter ablation of intraatrial reentry tachycardia is about 70% with a negligible complication rate. There are no reports of the use of radiofrequency ablation to treat other types of supraventricular tachycardia.

METHODS Standard diagnostic criteria were used to determine supraventricular tachycardia type. Appropriate sites for attempted ablation included 1) intraatrial reentry tachycardia: presence of concealed entrainment with a postpacing interval similar to tachycardia cycle length; 2) focal atrial tachycardia: a P-A interval ≤ -20 ms; and 3) typical variety of atrioventricular (AV) node reentry tachycardia: combined electrographic and radiographic features.

RESULTS Nine Mustard and two Senning patients underwent 13 studies to successfully ablate all supraventricular tachycardia substrates in eight (73%) patients. Eight of eleven (73%) patients having intraatrial reentry tachycardia, 3/3 having typical AV node reentry tachycardia, and 2/2 having focal atrial reentry tachycardia were successfully ablated. Among five patients having intraatrial reentry tachycardia (IART) and not having ventriculoatrial (V-A) conduction, two suffered high-grade AV block when ablation of the systemic venous portion of the medial tricuspid valve/inferior vena cava isthmus was attempted.

CONCLUSIONS Radiofrequency catheter ablation can be effectively and safely performed for certain supraventricular tachycardia types in addition to intraatrial reentry. A novel catheter course is required for slow pathway modification. High-grade AV block is a potential risk of lesions placed in the systemic venous medial isthmus. (J Am Coll Cardiol 2000;35:428–41) © 2000 by the American College of Cardiology

The Mustard and Senning operations were developed as surgical treatments for transposition of the great arteries. These operations fell into disfavor in part owing to a high incidence of arrhythmias at long-term follow-up. Intraatrial rentry tachycardia (IART) has been reported to occur in 2% to 10% of Mustard and Senning patients at 4 to 10 years' follow-up (1–5). Sudden cardiac death has been reported to occur late after these operations in 3% to 15% of patients (1,2,6,7), and a rapid ventricular response to paroxysmal atrial tachycardias has been thought to be the cause of death in many instances. Such morbidity and mortality has prompted an aggressive treatment philosophy. Older therapies have been far from perfect; antiarrhythmic drugs are fraught with proarrhythmic and noncardiac complications and with incomplete efficacy; antitachycardia pacing is only effective for slower tachycardias and may accelerate them to more dangerous, faster ones; and bradycardia pacing is adjunctive therapy, at best (8).

Based upon the high efficacy of radiofrequency catheter ablation to treat adults with typical atrial flutter (9,10), several investigators have reported a favorable early experience in Mustard and Senning patients having IART (11–14). These researchers utilized principles of concealed entrainment mapping (11–13), identification of nonconducting anatomic barriers (11,14) and presence of split potentials (13) to ablate IART circuits successfully in 70% to 80% of patients (11–13).

From the Departments of *Pediatrics, †Surgery, and ‡Internal Medicine, Duke University School of Medicine, Durham, North Carolina; the §Department of Pediatric Cardiology, Onassis Cardiac Surgery Center, Athens, Greece; and the ‖Department of Internal Medicine, University of North Carolina at Chapel Hill School of Medicine, Chapel Hill, North Carolina.

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Abbreviations and Acronyms

AV = atrioventricular
FAT = focal atrial tachycardia
IART = intraatrial reentry tachycardia
V-A = ventriculoatrial

Although these reports only described experience with IART, other supraventricular tachycardias may occur in patients with transposition. This report presents a single institution's experience with radiofrequency catheter ablation for various supraventricular tachycardias in patients who had undergone Mustard or Senning operations.

METHODS

Patient selection. All patients referred to Duke University Medical Center from January 1994 to March 1998 for electrophysiologic study and radiofrequency catheter ablation following Mustard or Senning operations were included (n = 10). An eleventh patient (patient 10) underwent his procedure at the University of North Carolina Memorial Hospital, where the first author (R.J.K.) participated. Patient demographic, surgical, and clinical arrhythmia data appear in Table 1. Supraventricular tachycardia had been documented in all patients, except patient 9, in whom the history suggested the diagnosis. Informed consent was obtained using a protocol approved by the Duke University Institutional Review Board for the first seven patients. A standard consent for electrophysiologic study and catheter ablation was obtained from the last four patients.

Anatomic descriptions. All operative reports were reviewed by a pediatric cardiac surgeon (R.M.U.) with attention to the locations of all incisions and the ultimate location of the coronary sinus with respect to the inferior suture line of the atrial baffle. Specific handling of the coronary sinus was described for 10 of the 11 patients (Fig. 1). If the baffle suture line kept the coronary sinus ostium in the systemic venous atrium, the medial isthmus between the tricuspid valve annulus and inferior vena cava was necessarily bisected, perhaps with the majority on the systemic venous side (Fig. 1c). Superiorly, this suture line always deviates from the tricuspid annulus to avoid the atrioventricular (AV) node. Hence, at least part of the medial isthmus would remain on the pulmonary venous side. If the coronary sinus was incised or other efforts made to redirect the ostium into the systemic venous atrium surgically, it was concluded that the great majority of the isthmus remained in the pulmonary venous atrium (Fig. 1b). Likewise, if the ostium was kept in the pulmonary venous atrium, the medial isthmus remained there as well (Fig. 1a).

The isthmus between the tricuspid valve annulus and the inferior vena cava orifice was arbitrarily divided into medial, mid, and lateral portions. If the tricuspid annulus were the face of a clock in a left anterior oblique projection, the anterior aspect of the medial isthmus would range from 4:00 to 5:00 o'clock, the mid isthmus from 5:00 to 7:00 o'clock, and the lateral isthmus from 7:00 to 8:00 o'clock, with each portion extending as a wedge posteriorly to the inferior vena cava orifice. The relationship of the isthmus subdivisions to the eustachian ridge was not considered, because the operative reports never mentioned this structure.

Definitions of tachycardia types. Tachycardia types were determined by a combination of surface electrocardiographic features and intracardiac electrophysiologic characteristics.

Intraatrial reentry tachycardia was paroxysmal in onset and termination and had a regular nonsinus P-wave morphology. It was inducible by atrial programmed stimulation or atrial burst pacing and had periods of second-degree AV block with a constant atrial cycle length. The mechanism of macroreentry was supported by demonstration of manifest entrainment.

Focal atrial tachycardia (FAT) was different from IART in its surface electrocardiographic appearance and intracardiac electrogram features. The P waves were discrete, with a clear isoelectric inter-P-wave interval in all leads. Split and fractionated electrograms were not present in regions of earliest atrial activation, including successful ablation sites. Unlike IART, manifest entrainment could not be demonstrated.

Typical variety of AV node reentry tachycardia was diagnosed when a regular, narrow-complex tachycardia could be induced by atrial or ventricular programmed stimulation in association with a 1:1 AV relationship, a ventriculoatrial (V-A) interval $\leq$70 ms, and concentric atrial activation. Because a His bundle recording could not be consistently obtained, standard maneuvers to dissociate ventricular or atrial electrograms from the tachycardia circuit were not possible. Additional findings supporting the diagnosis included 1) initiation of tachycardia during atrial extrastimulus testing following an AV interval increase of $\geq$50 ms during a decrease in atrial prematurity of 10 ms; 2) the observation that a single atrial extrastimulus initiates tachycardia following a double ventricular response; or 3) the ability of adenosine to terminate tachycardia following an atrial electrogram.

Technique of electrophysiologic study. Antiarrhythmic medications were discontinued at least four half-lives before each study. Intravenous propofol was used as a general anesthetic agent (15), except in Patients 2 and 6, who were studied under conscious sedation. Hemodynamic measurements were obtained in all patients, except Patients 6 and 10, in whom echocardiographic evaluation was performed. Diagnostic electrode catheters with close spacing (2 to 10 mm) were placed in the left ventricle, systemic venous atrium, and the inferior portion of the systemic venous remnant of the atrial septum in the region of the coronary

Table 1. Patient Demographics and Surgical and Arrhythmia History

Patient	Gender	Type of Atrial Switch	Age at Atrial Switch (months)	Location of Medial Isthmus From Figure 1	Other Cardiac Diagnoses	Prior Operations	Clinical Arrhythmias (Method of Documentation)	Failed Antiarrhythmic Drugs	Duration From First Tachycardia Symptoms to RFA (yrs)
1	M	Mustard	76	1b	VSD; severe PS	BAS (1 day); BTS (4 mos); VSD closure and LV-PA conduit (6 yrs) with Mustard	IART (ER visit)	Dg	7
2	M	Mustard	25	1a	None	BAS (1 day); patch enlargement of rt. atriotomy (2 yrs) with Mustard; VVI pacer for SAND (4 yrs); superior and inferior baffle stents for severe bicaval obstruction (21 yrs)	SAND with junctional bradycardia (ECG; Holter) IART (transtelephonic-pacer evaluation)	Q; Pr	3
3	F	Mustard	26	1b	VSD	BTS, BHS, and PAB (5 wks); VSD closure (2 yrs) with Mustard; VVI pacer for SAND (17 yrs)	SAND with junctional bradycardia (ECG; Holter) IART (ER visit)	Q; Di	19
4	M	Mustard	7	1c	VSD	BAS (1 day); VSA resection and patch closure of VSD (7 mos) with Mustard	IART (treadmill)	Dg	0.2
5	M	Mustard	6	1c	PDA	BAS (1 day); repeat BAS (3 mos); PDA ligation (6 mos) with Mustard	Nonsustained VT (Holter) IART (Holter)	None	1.5
6	F	Mustard	39	1b	None	BAS (1 day); VVI Pacer for SAND (10 yrs)	SAND with junctional bradycardia (ECG; Holter) IART (ER visit)	Di; Dg; At	1
7	F	Mustard	10	ND	Severe sub-PS	BAS (1 day); LV-PA conduit (7 yrs)	IART (Holter)	Dg; Q; V; Di	5
8	F	Mustard	7	1b	None	BAS (18 days); inferior baffle revision (4 yrs)	IART (Holter)	Dg	0.3
9	M	Senning	2	1d	None	BAS (1 day)	Palpitations (Hx only)	None	0.2
10	M	Mustard	7	1a	VSD	BAS (1 day); VSD closure (7 mos) with Mustard	AVNRT (prior EPS)	Dg; At	4
11	M	Senning	13	1d	VSD; severe sub-PS (unrepaired)	BAS (1 day)	IART (prior EPS)	None	0.7
Mean ± SD			19.8 ± 21.8 mos						3.8 ± 5.5 yrs

At = atenolol; AVNRT = AV node reentry tachycardia; BAS = balloon atrial septostomy; BHS = Blalock-Hanlon shunt; BTS = Blalock-Taussig shunt; Dg = digoxin; Di = diltiazem; EPS = electrophysiology study; ER = emergency room; Hx = history; IART = intraatrial reentry tachycardia; LV = left ventricle; ND = not described in operative note; PA = pulmonary artery; PAB = pulmonary artery band; PDA = patent ductus arteriosus; Pr = propranolol; PS = pulmonic stenosis; PVA = pulmonary venous atrium; Q = quinidine; RFA = radiofrequency ablation; SAND = sinoatrial node dysfunction; SVA = systemic venous atrium; V = verapamil; VSA = ventricular septal aneurysm; VSD = ventricular septal defect; * = isthmus between the tricuspid valve and orifice of inferior vena cava.

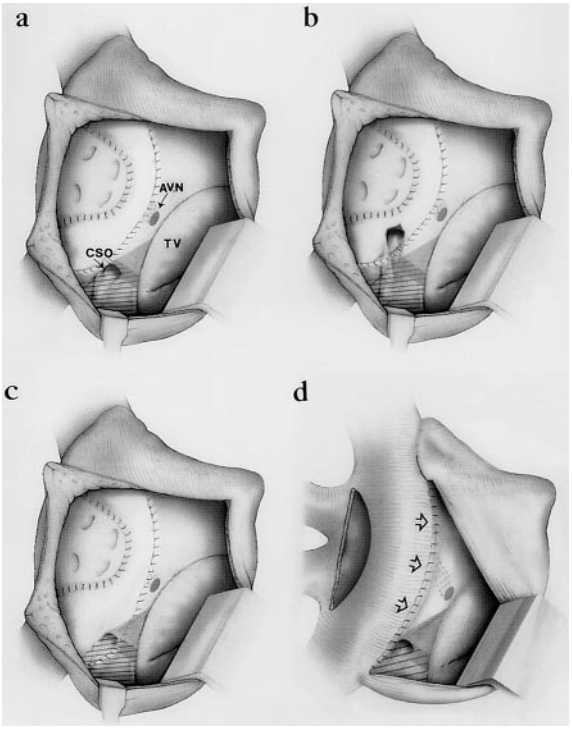

Figure 1. Diagrams of the pulmonary venous atrium following Mustard **(a–c)** and Senning **(d)** operations as viewed from a right anterior oblique projection. They illustrate the variations of surgical handling of the coronary sinus and medial isthmus between the inferior vena cava orifice and the tricuspid valve. **(a)** Inferior atrial baffle suture line is placed posterior to the coronary sinus ostium, keeping it and nearly all of the medial isthmus on the pulmonary venous side (Patients 2, 10). **(b)** Suture line is in approximately the same location as in **(a)**, but the ostium is surgically redirected posteriorly into the systemic venous atrium (Patients 1, 3, 6, 8). **(c)** Suture line is placed anterior to the coronary sinus ostium, keeping it and a large portion of the medial isthmus on the systemic venous side (Patients 4, 5). **(d)** Right atrial freewall edge **(arrows)**, which serves as a roof over the systemic venous tube, is sutured posterior to the coronary sinus ostium, similar in location to **(a)** (Patients 9, 11). AVN = compact AV node; CSO = coronary sinus ostium; TV = septal leaflet of tricuspid valve; **cross-hatched area** = medial isthmus; **shaded area** = region of "slow" inputs into AV node; **stippled area** = region of "fast" inputs into AV node.

sinus ostium. A His bundle recording was only obtained during ablation for AV node reentry tachycardia. A fourth diagnostic catheter, consisting of 5 to 10 bipoles, was placed within the systemic venous atrium from the inferior to the superior vena cava. In the Mustard patients, signals from the inferior-most bipoles recorded signals from the medial isthmus or right side of the inferior remnant of the atrial septum, the middle bipoles from the posterior wall of the native left atrium, and the superior-most bipoles from the right side of the superior remnant of the atrial septum (Fig. 2). In the two Senning patients, this catheter mapped the posterior length of the systemic venous tube—inferiorly, from the right side of the atrial septal remnant (due to surgical septectomy) adjacent to the IVC; then along the atrial septal flap; and superiorly, to the right side of the atrial septal remnant adjacent to the superior vena cava (Fig. 2). Patient 11 required transhepatic venous access, because there was bilateral iliofemoral venous thrombosis. Each patient was heparinized and the activated clotting time maintained at 250 to 300 s.

Standard pacing maneuvers were performed at baseline and under the influence of isoproterenol at 0.03 to 0.07 μg/kg/min. A 4-mm tip ablation catheter (EP Technologies, Sunnyvale, California; or Cordis-Webster, Baldwin Park, California) was then used for mapping purposes. Surface lead gains were increased to permit identification of the P-wave onset, and atria were mapped during tachycardia for presystolic activity and for split potentials. When 1:1 or 2:1 AV conduction was present, adenosine was used to identify P-wave onset. Regions in which there was electrical activity preceding P-wave onset were paced at cycle lengths 20 to 40 ms shorter than the atrial tachycardia cycle length to determine the presence of concealed entrainment.

Selection of sites for radiofrequency ablation. IART. An isthmus of atrial tissue was selected for creation of linear lesions based upon electrophysiologic characteristics and knowledge of the adjacent lines of conduction block, both natural and surgical. The presence of atrial presystolic activity was felt to be necessary, and was accompanied by split potentials if the site was adjacent to a suture line. Entrainment with concealed fusion proved that a site was critical to the IART circuit, if the atrial activation sequence was identical to that of tachycardia and if the postpacing interval was ≤30 ms longer than the IART cycle length (13). Natural barriers to conduction included the superior and inferior vena cava orifices and the tricuspid and mitral valve annuli. In Mustard patients, the baffle suture line and the right atriotomy were also potential lines of conduction block (Fig. 2). In Senning patients, the surgical anatomy is more complex, and there were several potential surgical lines of conduction block: 1) the suture line where the atrial septal flap forms a roof over the pulmonary veins; 2) the edge of the atrial septal remnant; 3) the suture line where the right atrial freewall attaches to the atrial septal remnant to form the roof over the systemic venous tube (Fig. 2, bottom: A–C); and 4) the suture line where the rightward most section of the pulmonary venous atrium is attached to the remaining right atrial freewall (Fig. 2, bottom: D–F).

FAT. Foci of earliest atrial activation were sought. Based upon published data from patients having atrial ectopic tachycardia (16), sites activated at least 20 ms prior to the P-wave onset were considered starting points for detailed mapping.

In cases of IART and FAT, radiofrequency ablation was considered successful if energy delivery during the tachycardia terminated the tachycardia, and if it could not be

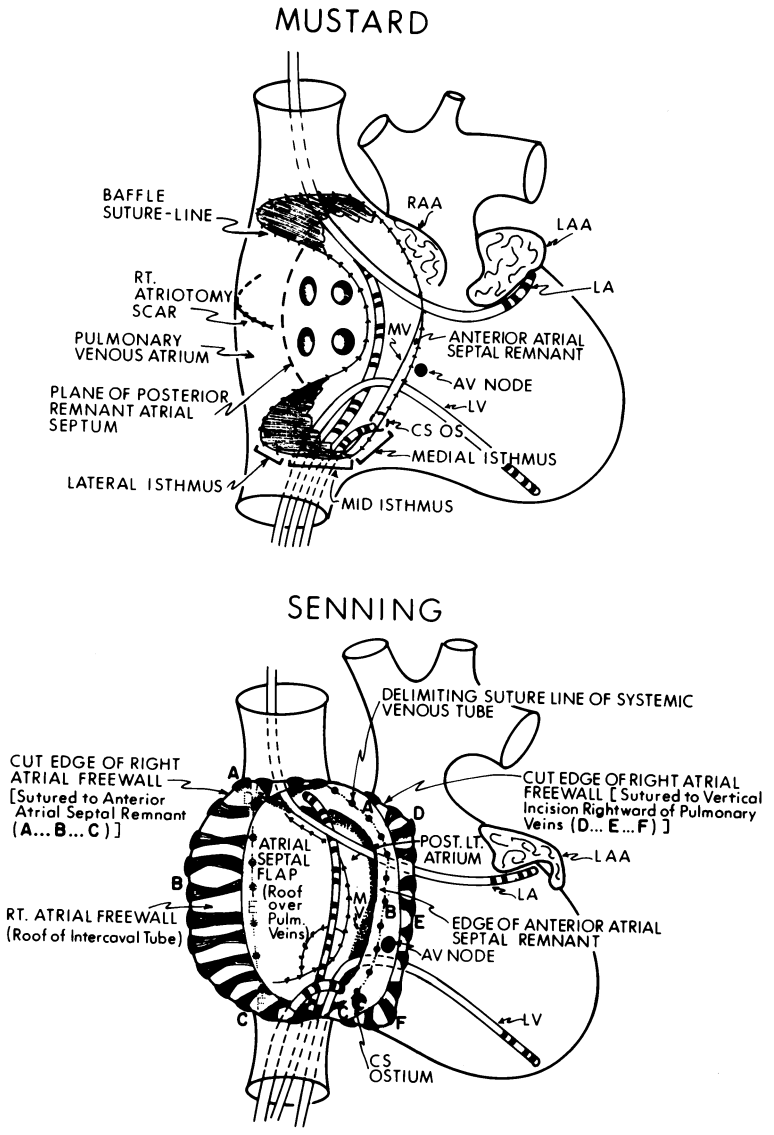

Figure 2. Diagrams of surgical anatomy with suture lines and of typical electrode catheter placement in Mustard and Senning patients. All important structures posterior to the level of the tricuspid valve are shown. The pulmonary venous atrium and the systemic venous baffle (cutaway) are illustrated in the Mustard diagram. The circle representing the coronary sinus ostium (CS os) is broken to emphasize its inconstant relationship with the adjacent portion of the baffle's suture line. In the Senning diagram, only the systemic venous atrium and the anterior atrial septal remnant portion of the pulmonary venous atrium are shown. See text for details. LA = left atrial catheter; LAA = left atrial appendage; LV = left ventricular catheter; MV = direction of mitral valve location; RAA = right atrial appendage.

reinduced for at least 60 min, at baseline and under the influence of isoproterenol.

ATRIOVENTRICULAR NODE REENTRY TACHYCARDIA. Radiofrequency modification of the slow inputs to the AV node was attempted using established principles published from patients having normal hearts (17,18). Access to the tricuspid annulus was possible only using an arterial catheter, which was retroflexed across the tricuspid valve into the pulmonary venous atrium. The catheter was pulled back and rotated clockwise until there was a dominant ventricular signal. Subtle flexion or extension was performed until a His bundle electrogram was obtained (Fig. 3, left). The catheter was further extended inferiorly until this electrogram disappeared (Fig. 3, right). Radiofrequency energy was delivered at this site during sinus rhythm. Accelerated junctional rhythm was considered a sensitive indicator of the appropriateness of the ablation site, and inability to reinduce AV node reentry tachycardia after 60 min was considered a specific indicator of successful ablation.

Statistical analysis. Descriptive data are presented as the mean ± SD, or, when appropriate, with 95% confidence intervals (CI). The Fisher exact test was used to analyze data on a nominal scale. Data obtained on a continuous scale

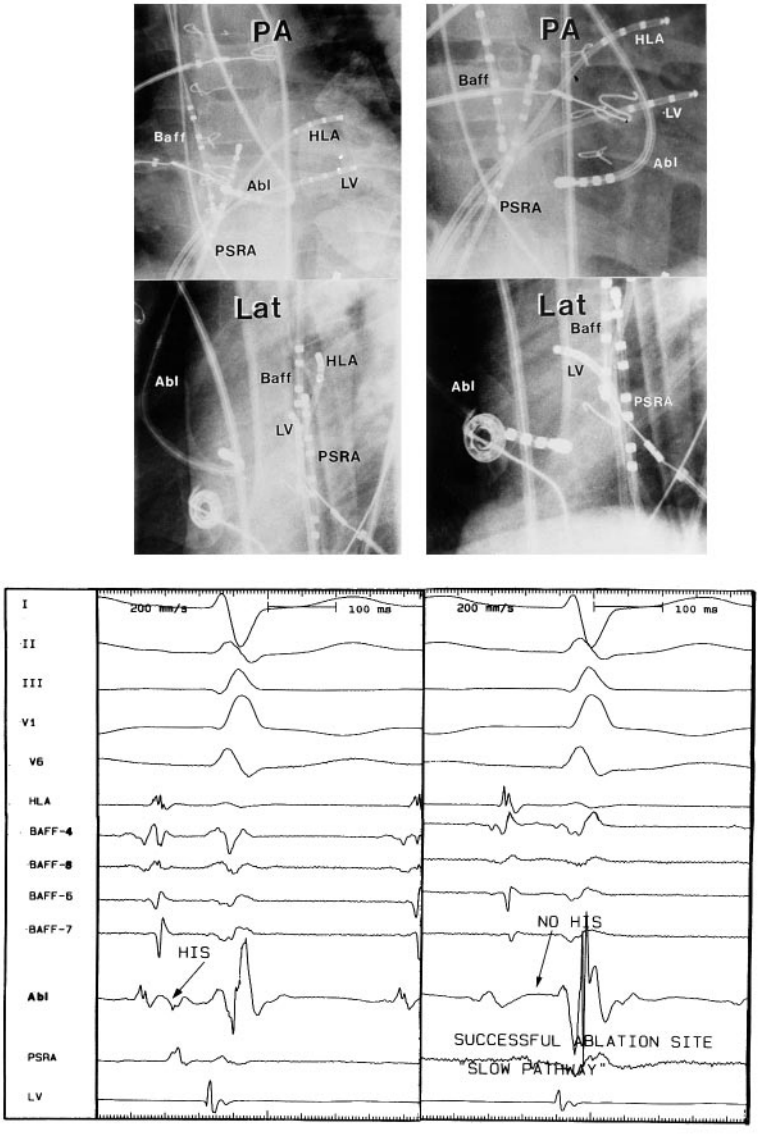

Figure 3. Posteroanterior (PA) and lateral (Lat) radiographs and electrograms during mapping for and ablation of the typical variety of AV node reentry tachycardia in Patient 9. **(Bottom Left)** The ablation catheter (Abl) was first used to identify the region of the His bundle using the retrograde aortic approach. **(Bottom Right)** The successful ablation site (Abl) of the "slow" inputs to the AV node. Radiographs on the left are lower magnification than those on the right. Baff = systemic venous baffle multipole catheter. Bipole 4 is most superior and bipole 7 most inferior; HLA = high anatomic left atrial catheter; LV = left ventricular catheter; PSRA = posteroseptal right atrial catheter. The electrograms are from the most proximal bipole. See text for details.

were analyzed using the Student *t* test. A p value <0.05 was considered statistically significant.

RESULTS

Patient characteristics. Eleven patients, ranging in age from 11 to 29 years, underwent 13 electrophysiologic procedures 10 to 27 years following their atrial switch operation (Tables 1 and 2). They had been symptomatic for two months to 19 years. Two patients had experienced syncope, two patients had symptomatic sustained IART requiring cardioversion, five patients had symptomatic non-sustained palpitations, and two patients had asymptomatic IART on Holter monitoring.

The patients had undergone Mustard or Senning operations ranging from 2 to 76 months of age (Table 1). Based upon the operative notes, the medial portion of the isthmus of tissue between the tricuspid valve and inferior vena caval orifice was almost exclusively left in the pulmonary venous atrium in eight patients (Fig. 1a,b,d) and was bisected between systemic and pulmonary venous atria in two patients (Fig. 1c). In patient 7 the operative note did not provide sufficient detail to determine this information. Six patients had coexisting structural cardiac defects, which are

Table 2. Cardiac Catheterization and Electrophysiology Study Results

Patient	Age at Study (yrs)	Hemodynamic Abnormalities	Baseline Rhythm	AVBCL (ms)	VABCL (ms)	Tachycardias Induced	Cycle Length (ms)
1	16	Complete SVC obstruction; small VSD; mild LVOTO (20 mm Hg)	Normal sinus	420	360 (iso)	IART	280
2	23	Mild bicaval obstruction (3–4 mm Hg)	VVIR paced	290	No conduction	IART	255
3	29	None	VVIR paced	400	No conduction	IART	245
4	11	Mild LVOTO (32 mm Hg); RV dysfunction (RVEDP = 12)	Normal sinus	340	No conduction	IART	220
5	15	None	Normal sinus	440	No conduction	IART	250
6	29	Hemodynamic study not performed	VVIR paced	430	No conduction	FAT (iso)	320
						IART #1 (iso)	215
						IART #2	300
	29	Hemodynamic study not performed	VVIR paced	280	390	IART #2	310
7	16	Mild LVOTO (30 mm Hg)	Normal sinus	300	No conduction	FAT (iso)	405
	18	Mild LVOTO (30 mm Hg)	Normal sinus	440	No conduction	IART	240
8	17	None	Normal sinus	410	620 (300 with iso)	IART	235
						Typical AVNRT	330
9	11	Mild superior baffle obstruction (2 mm Hg)	Junctional at 55 beats/min	260	690	IART	245
						Typical AVNRT	310
10	21	Hemodynamic study not performed	Junctional at 60 beats/min	400	500	Typical AVNRT	520
						IART (iso)	250
11	13	Unrestrictive VSD; arterial O_2 saturation—91%; severe LVOTO (80 mm Hg)	Normal sinus	330	670	IART #1	230
						IART #2 (iso)	200
Mean ± SD	19.0 ± 6.6 yrs			365 ms (95% CI = 328–402)			IART = 244 ms (95% CI = 229–257) FAT = 365 ms (95% CI = 280–446) AVNRT = 387 ms (95% CI = 256–518)

AVBCL = atrioventricular block cycle length; AVNRT = AV node reentry tachycardia; FAT = focal atrial tachycardia; IART = intraatrial reentry tachycardia; iso = isoproterenol; LVOTO = left ventricular outflow tract obstruction; RVEDP = right ventricular end-diastolic pressure; SVC = superior vena cava; VABCL = ventriculoatrial block cycle length; VSD = ventricular septal defect.

listed in Table 2. Three patients had undergone rate-adaptive, single chamber, ventricular pacing for "sick sinus syndrome," and none had undergone implantation of an atrial pacing lead.

Electrophysiologic study and radiofrequency ablation. Baseline hemodynamic and electrophysiologic data appear in Table 2. Antegrade AV conduction was present and decremental in every case. Because a stable His bundle recording was not obtained in every patient, the anatomic location of AV and V-A block was not always available. Ventriculoatrial conduction was present in six of the eight patients in whom the medial isthmus was known to be mostly on the pulmonary venous side, but was absent in the two patients in whom the native coronary sinus ostium was left on the systemic venous side (p = NS). Atrioventricular node reentry tachycardia was not inducible in any patient lacking V-A conduction. It was inducible in three patients having V-A conduction (50%) (p = NS).

IART. This tachycardia was inducible in all patients, and two patients had two apparently different circuits. Their mean cycle length was 244 ± 13 ms (range: 200–280 ms). A tachycardia thought to represent a circuit reversal of a previously identified circuit was counted as a single tachycardia. Radiofrequency energy was applied to the so-called critical zones of conduction as a series of stepwise connected burns creating linear lesions. Focal lesions were never successful for interrupting any IART. Overall, IART circuits were successfully ablated in 8 of 11 (73%) patients (95% CI for failure rate: 2% to 52%).

Mustard patients. Among nine Mustard patients, 10 apparently different IART circuits were identified. The isthmus of tissue between the tricuspid valve and inferior vena caval orifice was identified as a critical zone of conduction for nine of these circuits, and the region between the orifice of the superior vena cava and the superoposterior remnant of the atrial septum was a critical zone for one circuit (Fig. 4). In all five patients in whom it was attempted, concealed entrainment with a short postpacing interval could be demonstrated on both systemic and pulmonary venous sides of the inferior baffle.

All IART circuits were successfully ablated in six (67%) patients (95% CI for failure rate: 2% to 64%). Of the nine circuits involving the isthmus between the tricuspid valve and inferior vena caval orifice, only the medial isthmus could be negotiated from the systemic venous side due to the surgical anatomy; the inferior/anterior portion of the baffle always seemed to be very near the inferior vena caval orifice, leaving the entire mid- and lateral isthmus on the pulmonary venous side. Linear lesions were always attempted from the systemic venous side first, except in patient 2, due to the presence of an intravascular stent. In the other eight, this approach was successful in one, abandoned in two because of increasing AV block, and unsuccessful in five. When the systemic venous approach was unsuccessful, the retrograde, right ventricular approach to this region required prolapsing the flexed catheter across the tricuspid valve into the pulmonary venous atrium. Interruption of IART was usually only accomplished once the catheter tip was pulled back to the tricuspid valve annulus. In Patient 10, interruption of the IART circuit required that the linear lesion be extended more posteriorly to what was probably the right atriotomy scar. These pullbacks engaged the mid- or lateral isthmus, well away from the medial isthmus.

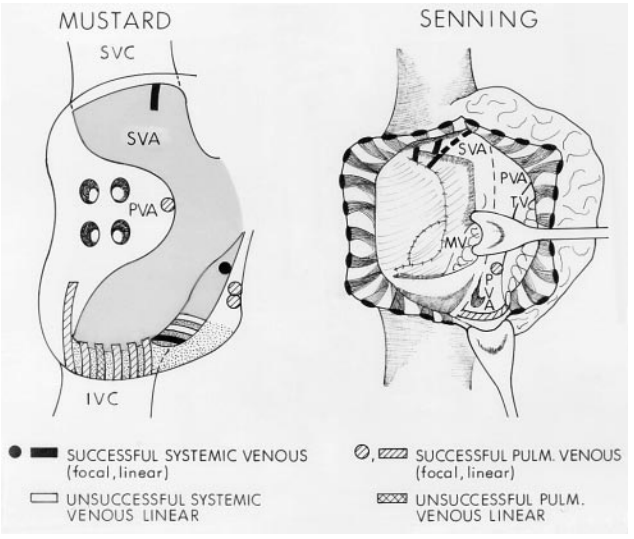

Figure 4. Locations of radiofrequency ablation attempts in Mustard and Senning patients. Illustrated are lesion locations for supraventricular tachycardias successfully and unsuccessfully ablated. In patients whose tachycardias were successfully ablated, unsuccessful locations are not illustrated. In the Mustard diagram, the **solid gray** area represents the systemic venous atrium, and the **stippled** area, the isthmus between the tricuspid valve and inferior vena cava. In the Senning diagram, the systemic venous atrium is opened, approximating the anatomy from Figure 2. The right atrial freewall flap comprising the roof over the systemic venous (intercaval) atrium is also removed, revealing the entire atrial septal remnant. The portion remaining on the pulmonary venous atrial (PVA) side includes the tricuspid valve/inferior vena cava isthmus. See text for details. MV = mitral valve; SVA = systemic venous atrium; TV = tricuspid valve.

Senning patients. The two Senning patients had three seemingly separate IART circuits (Fig. 4). In both patients, a critical zone of conduction was identified in the superoposterior remnant of the atrial septum below the superior vena caval orifice, but IART interruption did not occur until the linear lesions were continued inferiorly and posteriorly to the uppermost suture line where the atrial septal flap forms the roof over the pulmonary veins (Fig. 4). The catheter could be felt to "hop" over the cut edge of the atrial septal remnant to engage the more posterior structure. In patient 11, brief recurrences of this IART persisted until additional linear lesions were created posteriorly, from the superior-most portion of the native left atrium inferiorly to this same suture line (Fig. 4, broken black rectangle). The third IART circuit had as a critical zone of conduction the mid-isthmus, which had been entirely segregated from the

systemic venous atrium by the inferior portion of the superoinferior suture line attaching the right atrial freewall onto the inferior remnant of the atrial septum, posterior to the coronary sinus ostium (Fig. 4). Therefore, this could only be negotiated by using a retrograde arterial catheter and pulling back from this suture line to approximately "6 o'clock" of the tricuspid valve annulus. Both patients had successful ablation of IART circuits.

FAT. There were two FATs, each in Mustard patients (Tables 2 and 3). Both were inducible with atrial programmed stimulation. The cycle lengths of these tachycardias tended to be longer than those of the IARTs (363 ± 60 vs. 244 ± 13 ms, p = 0.2). The cycle length of the FAT from patient 7 varied from 360 to 450 ms and was markedly influenced by autonomic tone. It could also be terminated by adenosine. Sites of earliest atrial activation were in the pulmonary venous atrium in one and the systemic venous atrium in the other. Both foci were immediately adjacent to suture lines (Fig. 4) and were successfully ablated.

TYPICAL VARIETY OF AV NODE REENTRY TACHYCARDIA. This tachycardia was inducible in two Mustard patients and one Senning patient, but prior to delivery of radiofrequency energy only in patient 10. The cycle lengths of these tachycardias tended to be longer than those of the IARTs (387 ± 116 vs. 244 ± 13 ms, p = 0.16). The V-A intervals during tachycardia were 27 to 65 ms.

These three patients were successfully treated by applying radiofrequency energy along the tricuspid annulus below the location where a His bundle electrogram was observed (Figs. 3 and 4). Accelerated junctional rhythm was observed during radiofrequency energy in each. The PR intervals and the existence of V-A conduction were unaffected by these ablations.

Procedural issues and complications. These were lengthy procedures, in part due to the inclusion of hemodynamic studies. The total fluoroscopy times per patient (80 min; 95% CI: 62–99 min) and procedure times (8.8 h; 95% CI: 8.0 to 9.6 h) were longer than those reported from the Pediatric Radiofrequency Registry (19).

Two patients suffered second- or third-degree AV block during delivery of radiofrequency energy from the systemic venous side of the medial isthmus, and in one, it was permanent. These were the only two patients in whom the coronary sinus ostium was kept on the systemic venous side with no attempt at surgical redirection or incision of the coronary sinus (p = 0.02) (Fig. 1). These two patients were two of the five not having V-A conduction (p = NS). Among the eight patients whose medial isthmus was left almost exclusively on the pulmonary venous side, none suffered any degree of AV block (p = NS).

Follow-up. At 28 months' mean follow-up (range: 4 to 54 months), all 11 patients are alive (Table 4). Of those three whose ablations were not thought to be successful, one has a dual-chamber pacemaker for permanent second-degree atrioventricular block, and the other two receive class I antiarrhythmic drugs. All remain asymptomatic.

Among the eight patients whose ablations were thought to be successful, three receive digoxin, and one (Patient 10) atenolol. Digoxin had not been efficacious in those patients preprocedure. Patient 10 suffered IART recurrences, beginning two months following his procedure, but he is now completely controlled with atenolol. He had many recurrences prior to the procedure while receiving atenolol. Patient 4 has episodes lasting <3 s of a junctional tachycardia during exertion. All other patients remain asymptomatic. In summary, seven of the eight patients (88%; 95% CI for recurrence rate: 0% to 36%) whose procedures were successful have had no supraventricular tachycardia recurrences.

DISCUSSION

Radiofrequency ablation of FAT and atrioventricular node reentry tachycardia. The principal new information in this report is a description of radiofrequency catheter ablation of other supraventricular tachycardias in addition to IART following the Mustard and Senning operations. Sites of focal atrial tachycardias on either systemic or pulmonary venous sides of the surgical baffles can be mapped and ablated. These sites seemed to be immediately adjacent to baffle suture lines. Criteria previously described for accurate localization of atrial ectopic tachycardia foci (16) were useful in these two patients.

The typical variety of AV node reentry tachycardia was inducible in three patients; however, in two it was only seen following application of radiofrequency lesions to the medial isthmus for IART. These two cases could therefore have been caused by alteration of conduction properties of the slow inputs to the AV node, and, hence, could be iatrogenic. Irrespective of the etiology, application of radiofrequency energy along the tricuspid valve on the pulmonary venous side, probably corresponding to M1 and M2 zones (20), resulted in accelerated junctional rhythm (21) and rendered the tachycardia as noninducible. Well-established catheter ablation techniques (17,18,21) for the typical variety of AV node reentry tachycardia, then, proved efficacious in this patient group. The catheter course was similar to that required for isthmus ablation from the pulmonary venous side. However, more clockwise torque was required to render the tip medial and against the anteroseptal tricuspid annulus. To avoid damage to the compact AV node, we believe that it is critical to obtain a His bundle electrogram first. Flexion on the catheter may then be relaxed slightly until the His bundle signal no longer is present and there remain small atrial and large ventricular electrograms. In summary, we found both electrographic and fluoroscopic information to be necessary for safe and successful management of the typical variety of AV node reentry tachycardia.

Table 3. Radiofrequency Ablation of Supraventricular Tachycardias and Other Procedural Issues

Patient	Date of Study	Tachycardia	Criteria for Location of RF Energy Delivery	Successful (yes/no)	Main Site of Successful RFA	Main Site of Unsuccessful RFA	No. of RF Applications	Fluoroscopy Time (min)	Procedure Time (h)	Complications
1	1/94	IART	CEM; presystolic potentials	No		Medial isthmus (SVA linear; PVA focal)	23	110	11	Upper body edema (24 h)
2	8/94	IART	CEM; presystolic potentials	No		Mid- and lateral isthmus (PVA linear and focal)	7	50	9.5	None
3	3/95	IART	CEM; presystolic potentials	Yes	Medial isthmus (SVA linear)		10	35	8.5	None
4	12/95	IART	CEM; presystolic potentials	Yes	Mid-isthmus (PVA linear)	Medial isthmus (SVA linear)	20	65	9	Transient second-degree AV block
5	3/96	IART	Presystolic and split potentials	No		Medial isthmus (SVA linear)	3	40	8	Transient third-degree AV block; permanent second-degree AV block
6	2/96	FAT	Earliest atrial electrogram (A-P interval = 35 ms)	Yes	PVA-left of LPVs (focal)					None
		IART #1	CEM; presystolic potentials	Yes	SVC→septal remnant (SVA linear)		27	75	12	
		IART #2	CEM; presystolic potentials	Yes	Medial isthmus (SVA linear)					
	4/96	IART #2	CEM; presystolic potentials	Yes	Mid-isthmus (PVA linear)	Mid-isthmus (SVA linear)	11	40	8	None
7	4/94	FAT	Earliest atrial electrogram (A-P interval = 61 ms)	Yes	Mid-septal remnant (SVA focal)		17	75	10	None
	5/96	IART	CEM; presystolic potentials	Yes	Mid-isthmus (PVA linear)	Mid-isthmus (SVA linear)	8	35	6.5	None
8	11/96	IART	CEM; presystolic potentials	Yes	Mid-isthmus (PVA linear)	Medial isthmus (SVA linear)		111	8	None
		Typical AVNRT	Anatomic location of "slow inputs" to AV node; accelerated junctional rhythm	Yes	Mid-isthmus TVA (PVA focal)		20			

(continued)

Table 3. (continued)

9	5/97	IART	CEM; presystolic potentials	Yes	SVC→suture line of septal flap (SVA linear)	Mid-isthmus (PVA linear); medial isthmus (SVA linear)	17	88	7	None
		Typical AVNRT	Anatomic location of "slow inputs" to AV node; accelerated junctional rhythm	Yes	Mid-septal TVA (PVA focal)		10			
10	6/97	Typical AVNRT	Anatomic location of "slow inputs" to AV node; accelerated junctional rhythm	Yes	Mid-septal TVA (PVA focal)		7	102	8	None
		IART	CEM; presystolic potentials	Yes	Rt. Atriotomy scar→TVA (PVA linear)	Medial isthmus (SVA linear)	36			
11	3/98	IART #1	CEM; presystolic potentials	Yes	Lateral isthmus (PVA linear)	Mid-isthmus (SVA linear)	9	57	9	None
		IART #2	CEM; presystolic potentials	Yes	SVC/superior left atrium→suture line of septal flap (SVA linear)		8			
Mean ± SD							21/pt (95% CI: 14–28)	80 min/pt (95% CI: 62–99)	8.8 h/procedure (95% CI: 8.0–9.6)	

AVNRT = atrioventricular nodal reentry tachycardia; CEM = concealed entrainment mapping; FAT = focal atrial tachycardia; IART = intraatrial reentry tachycardia; LPVs = left pulmonary veins; PVA = pulmonary venous atrium; RFA = radiofrequency ablation; SVA = systemic venous atrium; SD = standard deviation; SVC = superior vena cava; TVA = tricuspid valve annulus.

Table 4. Current Patient Status at Last Follow-up

Patient	Duration Since Last RFA (mos)	Current Antiarrhythmic Medications	Current Rhythm Status	Clinical Tachycardia Recurrences
1	54	Dg	Normal sinus	None
2	47	Pr; Q	AAICP paced	None
3	40	None	VVIR paced	None
4	32	Dg	Normal sinus	None
5	28	Diso	DDD paced	None
6	27	None	VVIR paced	None
7	26	Dg	Normal sinus	None
8	20	None	Normal sinus	None
9	14	Dg	Junctional	Brief palpitation (<5 s runs, junctional tachycardia)
10	13	At	Junctional/sinus	IART
11	4	None	Normal sinus	None
Mean ± SD	28 ± 15 months (4–54 mos)			

At = atenolol; Dg = digoxin; Diso = disopyramide; IART = intraatrial reentry tachycardia; Pr = propranolol; Q = quinidine; RFA = radiofrequency ablation.

Radiofrequency ablation of IART. Our overall acute success rate for ablation of IART circuits (77% of circuits; 73% of patients) compares favorably with that described in previous reports (78% of circuits; 79% of patients) (11–13). Among Mustard patients having isthmus-dependent IART, Van Hare et al. (11) reported a relationship between the surgically determined location of the coronary sinus ostium and the location of successful radiofrequency ablation–pulmonary venous versus systemic venous side of the baffle suture line. When the ostium was on the systemic venous side, ablation on the same side was required for successful interruption of IART, and vice versa (11). Our findings did not agree with theirs.

Although the nomenclature in Table 1 is different from that used in the report by Van Hare et al., the "location of the medial isthmus" from Table 1 was similarly deduced from the description of the surgical handling of the coronary sinus. Whenever the coronary sinus was kept on the pulmonary venous side, or whenever it was surgically incised to permit redirection into the systemic venous atrium, we concluded that the medial isthmus was almost entirely pulmonary venous. Hence, a medial isthmus with both systemic and pulmonary venous components would correspond to a systemic venous coronary sinus in the Van Hare study (11). With that explanation, in only three of five Mustard patients in our series, in whom surgical procedural data were available, was the location of the coronary sinus predictive of the site of successful ablation IART; in three of four patients in whom the coronary sinus was left on the pulmonary venous side, ablation was successful on that side, having first been unsuccessful on the systemic venous side. In the other patient (Patient 3), ablation was successful from the systemic venous side. Among the two patients in whom the coronary sinus remained on the systemic venous side, in patient 4 the ablation was successful from the pulmonary venous side, having failed from the systemic venous side.

There are several possible explanations for the discrepancy in these findings. The small numbers of patients in both series make statistically based conclusions difficult. Second, with respect to the surgical handling of the coronary sinus, there may not be a fair comparison of patients in the two experiences. For example, in several patients in our series, we concluded that the medial isthmus was kept on the pulmonary venous side, when the coronary sinus ostium was surgically redirected to systemic venous atrium. If such surgical redirection was performed in the patients in the Van Hare series (11), then the expression "systemic venous coronary sinus ostium" would not have equivalence. Third, because of our experience with AV block in two patients during application of radiofrequency energy to the systemic venous side of the medial isthmus, we did not perform more than two linear lesions in this region in any subsequent patient before switching to the pulmonary venous approach. Hence, we became less aggressive.

Sites for delivery of radiofrequency ablation energy depended upon the tachycardia mechanism. For focal atrial tachycardia and AV node reentry tachycardia, the criteria used are discussed above. For IART, various maneuvers were used: 1) identification of fractionated atrial electrograms prior to the P-wave onset, preferably with split potentials, during tachycardia; 2) characterization of the anatomy based upon fluoroscopic location of the ablation catheter during mapping; and 3) proof of participation of that region in the tachycardia circuit, using entrainment with concealed fusion. In agreement with previous reports (11–13), when concealed entrainment was accompanied by

a postpacing interval ≤20 ms longer than the tachycardia cycle length, that location was considered to be a critical part of the tachycardia circuit. Knowledge of the anatomic boundaries of that portion of the circuit was then necessary to permit successful radiofrequency ablation.

The observation in Mustard patients that entrainment with concealed fusion and a short postpacing interval could be demonstrated on both systemic and pulmonary venous sides of the lower atrial baffle suggested to us that the entire isthmus participates in the reentrant circuit and must be transected. As the baffle extends anteriorly, to encircle the inferior vena caval orifice, catheter manipulation suggested that no portion of the mid- or lateral isthmus remained on the systemic venous side (Fig. 4). Except in patients in whom a large portion of the medial isthmus remains on the systemic venous side, the best opportunity for transecting the entire isthmus is from the pulmonary venous side at the level of the mid- or lateral isthmus.

AV block during radiofrequency ablation. This is the first report of damage to AV conduction during radiofrequency ablation of the medial isthmus from the systemic venous side following the Mustard procedure. To understand how this might have occurred, precise knowledge of the original surgical procedure is necessary. Essential components of the Mustard operation include a subtotal atrial septectomy followed by placement of a complex baffle of pericardium or prosthetic material, described as "pant legs" in shape. Damage to anterior or "fast" inputs to the AV node might be expected by the septectomy, although there are no intraoperative mapping data that precisely address this anatomic region. Electrocardiographic evidence for this would include first-degree AV block; however, prolongation of the PR interval is frequently seen after this procedure due to severe intraatrial conduction delay (22). Confounding our ability to discriminate the anatomic level of PR interval prolongation was an inability to obtain a reliable His recording. Based upon our knowledge of AV node conduction in the normal heart, we considered the presence of intact V-A conduction as suggestive of at least adequate function of the "fast" inputs to the AV node.

With respect to posterior or "slow" inputs to the AV node, conduction block to the entire posteroseptal region has been demonstrated during intraoperative mapping, when the coronary sinus was kept on the systemic venous side of the inferior baffle suture line (23). A portion of the "slow" inputs to the AV node are thought to exist in this region (Fig. 1). The two patients in our series suffering at least transient second- or third-degree AV block were among the six in whom the coronary sinus ostium drained into the systemic venous side and the only two in whom it had not been surgically redirected or incised. Therefore, the combination of the original surgical technique plus ablation catheter access to the medial isthmus from the systemic venous side (Fig. 1c) may have caused "slow pathway" damage. If the absence of V-A conduction observed in these two patients represents prior loss of "fast pathway" conduction, then overall impairment of AV node conduction might be expected.

Study limitations. There are several limitations of this study. Anatomic detail was inferred from the fluoroscopic location of catheters coupled with prior echocardiographic and angiographic information. Real-time transesophageal echocardiographic or intracardiac echographic (24) techniques would provide new and potentially more accurate information regarding anatomy and catheter contact. Mapping IART in such complex structures may also benefit from new magnetic electroanatomic mapping techniques such as those described by Marchlinski et al. (25) and by Dorostkar et al. (26). In our patients, no attempts were made to demonstrate creation of bidirectional block to conduction, either during or following radiofrequency energy delivery to IART circuits. This will require novel catheter placement in future patients. The absence of a reliable His bundle electrogram throughout all studies prevented meaningful evaluation of AV node function. This would have been most important in the two patients suffering AV node damage. Follow-up longer than 28 months will be necessary to determine the long-term efficacy of radiofrequency ablation for supraventricular tachycardias in these patients. In particular, it is hoped that the recurrence rate of IART will be as low as that seen in adults with normal hearts and not as high as that seen in Fontan patients. We speculate that the relatively normal atrial size and pressure in our patients—in distinction from that observed in Fontan patients—will predict this to be so.

Clinical implications. As the population of patients who had undergone the Mustard or Senning operation ages, paroxysmal tachycardias remain their most significant clinical problem. The majority of these tachycardias are supraventricular in origin, predominantly IART. Other than atrial fibrillation, there appears to be no supraventricular tachycardia that should be considered exempt from candidacy for radiofrequency ablation following these operations. As in patients having normal cardiac anatomy and common atrial flutter, risk of damage to the AV node should be discussed with patients, as ablation of inputs to the AV node may occur.

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Reprint requests and correspondence: Dr. Ronald J. Kanter, Duke University Medical Center, Box 3090, Durham, NC 27710. E-mail: kante001@mc.duke.edu.

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