CASE REPORTS

Cryoablation of Drug-Resistant Ventricular Tachycardia in a Patient with a Variant of Scleroderma

John J. Gallagher, M.D., Robert W. Anderson, M.D., Jackie Kasell, John R. Rice, M.D., Edward L. C. Pritchett, M.D., James H. Gault, M.D., Lura Harrison, Ph.D., and Andrew G. Wallace, M.D.

SUMMARY A 37-year-old man with a benign variant of scleroderma (CRST syndrome: calcinosis circumscripta, Raynaud's phenomenon, sclerodactyly, and telangiectasia) presented with recurrent ventricular tachycardia. Preoperative electrophysiologic study suggested that the mechanism of tachycardia was an ectopic pacemaker focus in the right ventricle. Right ventricular dilatation, tricuspid insufficiency, normal pulmonary pressures, and normal coronary arteries were also demonstrated. At surgery, epicardial mapping localized the site of origin of ventricular tachycardia to the anterior right ventricle near the crista supraventricularis. Intramural recordings of the site of tachycardia demonstrated autonomous activity unreflected on the peripheral ECG during brief periods of sinus rhythm. Local epicardial cooling of this area with a cryoprobe promptly terminated ventricular tachycardia with resumption of tachycardia on warming. The focus was ablated by freezing the area at -60°C. The patient remained free of dysrhythmia on no antiarrhythmic agents for eight months at which time he had a single recurrence of ventricular tachycardia from a different site in the right ventricle. This technique offers a method for ablating sites of dysrhythmia arising in diffusely diseased myocardium.

RECURRENT VENTRICULAR TACHYCARDIA resistant to conventional drug therapy and pacemaker techniques has been treated with surgery in a number of recent cases. Procedures previously reported with variable success have included: revascularization, sympathectomy, a cryosurgery, and replacement of the mitral valve in cases of balloon mitral valve syndrome.

The purpose of this report is to describe successful localization of the site of origin of ventricular tachycardia in a patient with CRST syndrome (calcinosis circumscripta, Raynaud's phenomenon, sclerodactyly and telangiectasia), a variant of scleroderma, and ablation of the focus using a cryosurgical technique.

Case Report

A 37-year-old Caucasian man was admitted in July 1976 to Duke University Medical Center with a 3½ month history of recurrent ventricular tachycardia refractory to conventional therapy.

During the 12-year period prior to onset of the rhythm disturbance the patient had experienced slowly progressive circumscribed calcinosis, cold-induced cyanosis and fingertip ulcerations involving the digits of both hands. Additional subcutaneous calcium deposits developed over both olecranon areas, over the left Achilles tendon and on the right small toe. Fingerpad telangiectases and patchy induration of the right forearm skin were first noticed during the two years prior to admission. Other than occasional difficulty with minor arthralgias there was no clinical history suggestive of major organ involvement. Specifically, there was no prior history of exercise intolerance, irregular pulse, hypertension, renal dysfunction, dyspnea, dysphagia or altered bowel habits. The general connective tissue disease review of systems was similarly unremarkable.

In March 1976, the patient abruptly developed substernal and abdominal discomfort associated with rapid heart rate. A standard electrocardiogram suggested ventricular tachycardia at a rate of 150 beats/min. The patient was admitted to his local hospital where he was cardioverted. Balloon flotation catheterization revealed an RA mean pressure of 16 mm Hg with "cv" waves of 19 mm; right ventricular pressures = 32/20 mm Hg; pulmonary artery pressures = 32/15 mm Hg with a mean of 20 mm Hg. The cardiac index was 2.6 L/min/m². Tricuspid insufficiency persisted in sinus rhythm and was attributed to primary right ventricular failure. Therapy with digoxin 0.25 mg daily and quinidine sulfate 300 mg every six hours maintained normal sinus rhythm. After a 5 week hospital course complicated by pulmonary embolism the patient was discharged on digoxin, quinidine and Coumadin, only to be readmitted in May 1976 with recurrence of the ventricular arrhythmia requiring cardioversion. Cardiac catheterization at that time demonstrated smoothly patent coronary arteries with a dominant left coronary system. Pressures determined at this time demonstrated right atrium: a = 16, v = 11, mean = 9; right ventricle: pulmonary artery pressures = 26/10, mean 15; mean pulmonary capillary wedge = 10; left ventricle = 117/0–13. The cardiac index was a 2.4 L/min/m² and the LV ejection fraction was 44%. Dosage of digoxin was decreased empirically to 0.125 mg daily and he was discharged on his previous regimen.
Three weeks later, in May of 1976, he was readmitted to his local hospital with a third episode of symptomatic tachycardia. A His bundle study confirmed the ventricular origin of the arrhythmia based on the findings of ventriculo-atrial dissociation and absence of His deflections before beats of ventricular tachycardia. Attempts at both atrial and ventricular overdrive pacing were ineffective. Right ventricular angiography documented an end-diastolic volume of 280 cc/m², a cardiac index of 1.65 L/min/m², and tricuspid regurgitation of roughly 30% of right ventricular volume. Coronary sinus pacing to a rate of 80 beats/min increased the cardiac index to 2.4 L/min/m² and a permanent transvenous pacing catheter was placed in the coronary sinus. In spite of continued drug therapy and fixed atrial pacing at 80 beats/min, ventricular tachycardia recurred. The combined use of propranolol 150 mg and quinidine sulfate 300 mg every six hours, in addition to digoxin 0.125 mg daily, was ineffective in restoring sinus rhythm. All drugs except digoxin were therefore discontinued and disopyramide phosphate 200 mg every six hours was begun. Sinus rhythm returned and the patient was discharged after 7 days with arrhythmia.

One month later, in June 1976, he was again admitted to his local hospital with recurrent ventricular tachycardia, this time at 120 beats/min. He was noted to have runs of ventricular tachycardia 4 hours after his dose of disopyramide every 4 hours. Tachycardia recurred and prompted therapy with quinidine sulfate 200 mg in addition to disopyramide phosphate 300 mg every six hours. Despite this, tachycardia recurred. Quinidine was discontinued and he was treated with propranolol 20 mg in combination with disopyramide phosphate 300 mg every six hours. He continued to have ventricular tachycardia at a rate of 100-120 beats/min. Propranolol was discontinued with no apparent effect. Despite increasing the rate of coronary sinus pacing to 116 beats/min, he remained in ventricular tachycardia for approximately 70% of every 24 hour period, and was therefore referred to Duke University Medical Center for consideration of possible surgical intervention.

Physical Examination

Physical examination revealed a chronically ill man in obvious cardiorespiratory distress. Vital signs on admission included a regular pulse rate of 120 beats/min, BP 110/80, respirations 22 beats/min, and temperature 37°C. The skin over the malar areas and right volar forearm was found to be somewhat indurated and “bound down.” Sclerodactyly with evidence of healed fingertip ulcerations was present on all upper extremity digits and hard subcutaneous deposits were present over the olecranon areas bilaterally. Telangiectases were noted over the distal aspects of the fingers and a single telangiectasia was discovered on the mucosal surface of the lower lip. Pigmentation was normal as were the hair and nails. The remainder of the head, eye, ear, nose and throat examination was otherwise normal as was examination of the chest and lungs.

Cardiovascular exam revealed elevated neck veins demonstrating “cv” waves, and an occasional “cannon” wave. The venous pressure was not elevated. The carotid pulses were normal. A diffuse precordial impulse was felt from the left sternal border extending 8 cm in the fifth left intercostal space. There was no sternal lift present, and no area of medial retraction could be identified in the precordial impulse. The first sound varied in intensity. Splitting of the second sound was difficult to evaluate in the presence of tachycardia, but there was no obvious accentuation of the pulmonic component. A soft grade II/VI systolic murmur beginning with the first sound and extending through half of systole was noted on inspiration. An S₃ was also audible which increased with inspiration.

Abdominal examination revealed a liver edge palpable 4–5 cm below the right costal margin, with a total span of 14 cm. The edge was firm, slightly tender, but nonpulsatile.

Laboratory Examination

The electrocardiogram on admission (fig. 1) demonstrated tachycardia at 150 beats/min associated with left bundle branch block configuration with a left axis deviation. Intermittent pacemaker artifact from the coronary sinus pacemaker was observed, resulting in ventricular capture with fusion. The chest X-ray demonstrated prominent enlargement of the right atrium and ventricle. An electrode catheter was positioned in the coronary sinus and the lung fields were clear.

Routine admission laboratory studies were essentially

![Figure 1. Electrocardiogram during ventricular tachycardia. Pacing spikes due to the coronary sinus pacemaker appear in the tracing. A wide QRS tachycardia with a rate of 150 beats/min is present associated with a configuration resembling left bundle branch block with leftward axis deviation. A-V dissociation is present.](http://circ.ahajournals.org/lookup/suppl/doi:10.1161/01.CIR.31.11.191/-/DC1/fig1.jpg)
normal. Serum creatinine was 1.3 mg%; Westergren erythrocyte sedimentation rate, 2 mm/hr; IgG FANA, positive 1:40 with a nucleolar pattern; RA factor by latex was negative; digoxin level was 1.1 ng/ml. Hand X-rays showed multiple areas of soft tissue calcification and tuft erosion of the fourth right finger.

Electrophysiologic Study

Electrophysiologic study was performed in the postabsorptive, nonsedated state. The patient was on no cardioactive medications and was in ventricular tachycardia with a rate of 140 beats/min on entering the laboratory. A #6 F Lehman catheter was introduced percutaneously via the right femoral vein and advanced to the right side of the heart. Tricuspid regurgitation was present. The pulmonary artery pressure was 20/12 with a mean of 15 mm Hg. The pulmonary capillary wedge pressure was 12 mm Hg. The cardiac index measured 1.8 L/min/m². This catheter was then replaced by a tripolar electrode catheter for recording the His bundle. In addition, catheters were placed in the right atrium and right ventricle. The implanted coro-

![Figure 2](image)

**Figure 2.** Intracavity recordings during ventricular tachycardia. Tracings from above down are standard ECG lead V1, I and bipolar electrograms recorded from the right atrium (RA), region of the His bundle (HBE) and the right ventricular apex (RV). S = stimulus artifact; A = atrial electrogram; H = His bundle; V = ventricular electrogram. At the beginning of the recording atrial pacing with overdrive of the tachycardia is present at a cycle length of 400 msec. A His deflection precedes each ventricular complex with a normal H-V interval. The atrial pacing is then discontinued and following the last conducted beat, ventricular tachycardia and A-V dissociation appears. Note the absence of His deflection preceding the wide QRS complexes of ventricular tachycardia.

![Figure 3](image)

**Figure 3.** Epicardial map during ventricular tachycardia. The isochronous lines of epicardial activation are referenced to the onset of the QRS complex as recorded by standard ECG leads (O time). The earliest area of epicardial activation occurs on the anterior right ventricle 85 msec before the onset of the surface ECG deflection.
nary sinus pacemaker was adjusted to cycle at a slower rate.

At the onset of the study, a wide anomalous QRS tachycardia was present with A-V dissociation at a ventricular cycle length of 400 msec. Atrial pacing at cycle length of 380 msec resulted in 1:1 A-V conduction over the normal conduction system, associated with an A-H interval of 180 msec, an H-V interval of 50 msec, and a QRS complex 0.08 sec in duration with loss of inferior wall forces. On termination of atrial pacing (fig. 2), ventricular tachycardia reappeared with no His bundle deflection preceding the wide QRS complexes. Further atrial pacing demonstrated the Wenckebach phenomenon proximal to the recorded His deflection at a cycle length of 360 msec.

Recordings made in the right ventricle (RV) using the RV cavity potential as a reference revealed the following activation times: RV-apex = 35 msec; RV-septum(HBE) = 50 msec; RV body of RV = 33 msec; RV-inflow = 80 msec; RV-outflow = 40 msec.

Premature ventricular beats were introduced synchronously throughout diastole after every eight spontaneous tachycardia beats either singly or in pairs. These premature beats demonstrated examples of penetration of the focus of tachycardia with reset; some premature beats appeared to induce exit block from the focus. Very early premature beats lengthened the return cycle length of tachycardia. No examples of shortening of the tachycardia cycle length were noted, and no combination of premature beats could terminate the tachycardia. An attempt was made to overdrive the tachycardia by ventricular pacing, and the time of reappearance of tachycardia on termination of pacing determined. The "escape time" at the cycle lengths (CL) tested were: CL 360 = 580–650 msec; CL 320 = 690 msec; CL 280 = 700 msec. The rhythm was always perfectly regular after termination of pacing at all rates.

Our conclusions from the electrophysiologic study were that ventricular tachycardia arising in the body of the right ventricle was present which appeared unifocal and due to an ectopic pacemaker. Because of the intractable nature of the arrhythmia, it was elected to attempt surgical ablation of the site of arrhythmia.

Operative Intervention

At surgery, a median sternotomy was performed and the pericardium was opened. The right atrium and ventricle appeared to be markedly dilated and the right ventricle contracted poorly. Multiple areas of scarring were noted but no discrete aneurysm was detected. The left atrium and left ventricle appeared to be normal. Cannulas were placed in the aorta and in both the inferior and superior vena cava. A reference needle electrode was inserted on the anterior right ventricle and epicardial mapping begun during ventricular

---

**Figure 4.** Electrograms recorded from the site of ventricular tachycardia. Recordings in panels A-F from above down are standard ECG leads I-III, a bipolar reference recorded from the right ventricular epicardium, and unipolar and bipolar recordings from a transmural needle electrode positioned at the site of origin of earliest epicardial activation during ventricular tachycardia. A ten millivolt calibration signal for the unipolar tracings appears in panel F.
transitions between ventricular tachycardia and sinus tachycardia were observed. These transitions generally occurred over a period of 15-20 heart beats. One such representative transition is illustrated in figure 4. Ventricular tachycardia was initially present in panel A and the activity recorded on the needle electrode occurred well before the onset of the surface ECG deflections. In panel B and C, the sinus P wave gradually emerged from the QRS resulting in fusion beats in panel C. Several beats later, in panel D, the QRS assumed the configuration present in sinus rhythm. Despite this, the electrode recording adjacent to the site of ventricular tachycardia continued to record activity unreflected by surface ECG. After approximately ten more beats, the activity recorded by the needle electrode gradually merged into the QRS (panel E) finally becoming "locked in" to the end of the QRS as shown in panel F. Electrograms recorded from the site of origin of ventricular tachycardia (fig. 5) during a period of sinus rhythm demonstrated a split complex with late activation occurring after the inscription of the peripheral QRS complex.

After the ectopic focus had been mapped in this manner, the patient was placed on cardiopulmonary bypass under normothermic conditions. Using the technique previously described, a cryoprobe (fig. 6) was placed on the epicardial surface at the site of ventricular tachycardia and cooling was commenced to 0 degrees. After 60 seconds the ventricular tachycardia promptly stopped. It recurrd some 10 sec after the cooling was discontinued. Three separate freezes were therefore carried out in this localized area, each at a temperature of -60 degrees C for a period of 90 sec. At this time the tachycardia had disappeared and the heart rate was 50. Outflow from the right ventricle was then re-established and the right atrium was exposed for further study (fig. 7).

*Fabricated to our design by Frigitronics.
The conclusion of this procedure no further evidence of ectopic activity appeared. No potentials of local activation could be recorded from the transmural needle. To ensure complete ablation of activity in the endocardial layers however, a superficial purse string suture was placed on the epicardial surface, and a cryoprobe fashioned in the form of a trochar (fig. 7) was passed through the ventricular wall. A final lesion was created with this probe again freezing for 90 sec at a temperature of −60 degrees C. The probe was then withdrawn and the purse string suture tied. The right atrium was briefly opened and the tricuspid valve was inspected, revealing no evidence of Ebstein's anomaly. The right ventricular wall was palpated and noted to be intact in the area of freezing. No abnormally thin areas could be detected.

Biopsies from the right atrium and pericardium and needle biopsies of the right and left ventricle were obtained. The patient was withdrawn from cardiopulmonary bypass and the incisions closed in the usual manner.

Postoperatively he did well and was discharged 10 days after surgery on no medication other than maintenance digoxin 0.25 mg daily.

Routine microscopy of the right and left ventricular biopsies showed unremarkable myocardium. A section of the right atrium showed an area of fibrin deposition within the endocardium. Marked fibrosis was observed in the pericardium. The right atrial myocardium, right ventricular myocardium, and pericardium were evaluated by immunofluorescence examination. There was no significant localization of IgG, IgM, IgA, C₃ or albumin within the section of right ventricle. The sections of right atrium showed no localization IgG, IgM, IgA, fibrinogen, or albumin. However, several foci of complement component C₅ were present in a clumped pattern in unidentifiable structures. Sections of a full thickness of pericardium showed no localization of IgG, IgM, IgA, fibrinogen, or albumin. Complement component C₅ localized along the endothelial lining of several venous channels in the pericardial biopsy. The localization of complement component C₅ in the absence of any other serum proteins examined was thought to be nonspecific.

The postoperative electrocardiograms (fig. 8) were identical to the few available preoperative tracings in sinus rhythm, demonstrating low voltage, loss of inferior wall forces and nonspecific ST-T wave changes.

Follow-up

Following discharge from the hospital, he remained free of arrhythmia on no antiarrhythmic agent. He developed occasional mild edema responding to diuretic therapy. His exercise tolerance gradually improved and he returned to gainful employment.

Eight months after surgical ablation of his site of ventricular tachycardia, he developed ventricular tachycardia. A 12 lead electrocardiogram was recorded, (fig. 9) demonstrating a QRS configuration different from all previously recorded arrhythmias. Cardioversion was performed with uneventful conversion to sinus rhythm and the patient was released on prophylactic quinidine sulfate 200 mg every 6 hours. On this regimen he has continued again free of arrhythmia.

Discussion

Surprisingly little is known about the exact substrate required for ventricular tachycardia to occur, but undoubtedly there is a variety of possible anatomic situations ranging from congenital or acquired disorders of the conduction system with grossly normal myocardium to frank aneurysm formation. Resection of aneurysm can be an effective treatment for recurrent ventricular tachycardia especially when epicardial mapping is used to localize the origin of dysrhythmia to the anatomic abnormality. The situation is less clear when there are large dyskinetic areas involved, especially when hemodynamic function is borderline. The surgical approach to a discrete zone of pathology situated near a critical area (e.g., base of a papillary muscle) or to a site of dysrhythmia with grossly normal myocardium is even
more uncertain. One interesting approach has been suggested by Guiraudon and Fontaine who advocate performing a transmural ventriculotomy through the site of dysrhythmia. The ventriculotomy and its subsequent surgical closure presumably interrupts potential re-entrant circuits situated in the Purkinje-myocardial junctions. The greatest success of this technique has been obtained in patients whose ventricular tachycardia does not result from ischemia and which exhibits characteristics of a re-entrant mechanism. Another approach used by Cournel in the atrium is the use of cautery. Cautery is irreversible, denatures collagen, and thus results in loss of tensile strength with a risk of rupture or aneurysm formation if applied to the ventricle.

The use of cryothermia offers the advantage that an initial reversible block can be induced to test a particular area in the area of dysrhythmia before proceeding to irreversible ablation. The size of the lesion can be easily titrated by temperature adjustment of the cryoprobe and monitoring of the boundary area with thermocouple devices. The chronic lesion which results is characteristically composed of dense homogeneous connective tissue. This approach had been previously used to ablate the A-V node-His bundle as well as accessory atrioventricular connections in patients with refractory arrhythmias. This report extends the method to ablation of ventricular tachycardia.

Cardiac involvement due to systemic sclerosis cannot be definitely implicated in the genesis of our patients arrhythmia but the association of ventricular dysrhythmia with a supposedly benign variant of scleroderma appears to merit further observation. Cardiac involvement in scleroderma has been noted by several authors and may involve the endocardium, myocardium, or pericardium — singly or in combination. The myocardium is most frequently involved and this generally consists of replacement of cardiac muscle by connective tissue. Diffuse patchy fibrosis of the entire right ventricle was apparent by gross inspection and epicardial mapping. The failure of the single needle biopsy of right ventricle to demonstrate this suggests that the area sampled was relatively spared (biopsy taken from apex). Our patient had no obvious pulmonary or renal involvement and appears to have rather selective primary involvement of the right ventricular myocardium. An alternative hypothesis was that he has the syndrome of “arrhythmogenic right ventricular dysplasia.” This entity has been recently proposed by Fontaine and our patient indeed exhibits many of its features. Arrhythmogenic right ventricular dysplasia is characterized by 1) “post-excitation” in sinus rhythm (e.g., activation of areas of myocardium after inscription of the QRS complexes as recorded by standard ECG leads); 2) the QRS morphology of ventricular tachycardia demonstrates left ventricular delay; 3) right ventricular angiography demonstrates areas of diffuse or discrete thinning of the ventricular wall usually located in the infundibulum of the right ventricle; 4) the mechanism of arrhythmia suggests re-entry. Our patient had “post-excitation” and origin of tachycardia near the infundibulum of the right ventricle was documented by epicardial mapping. Angiography however failed to show thinning of the right ventricle and this was not apparent at surgery. We of course can not exclude microreentry as the mechanism of dysrhythmia.

The electrocardiographic findings reported in scleroderma have varied from normal electrocardiograms to all varieties of conduction disturbances and hypertrophy. Arrhythmias have been reported but ventricular tachycardia is distinctly unusual.

The episode of ventricular tachycardia appearing in our patient eight months after surgery is not thought to be due to the original site of tachycardia based on analysis of the electrocardiogram but this event does emphasize the fact that progression of the underlying process is always a risk in chronic disease.

The technique of using cryothermia to ablate sites of rhythm disturbances requires further study. Since a rather discrete lesion is employed, localization of the site of

**Figure 9.** Electrocardiogram during recurrent ventricular tachycardia eight months after surgery. This electrocardiogram demonstrates a rapid ventricular tachycardia with left bundle branch block morphology. In contrast to the previous tachycardia, the axis of this tachycardia is different. In addition the precordial morphology in leads V₁-V₆ is different.
CLOSURE OF CORONARY FISTULA/Shubrooks, Naggar

197

Dysrhythmia must be very exact. With current methods, this limits application of the technique to cases where a stable unifocal ventricular tachycardia with a rate less than 200 beats/min is present in the operating room. It appears likely however that in the near future with the use of multiple simultaneously recorded electrodes and on-line computer analysis, more complex ventricular dysrhythmias may lend themselves to possible surgical intervention.

References

Spontaneous Near Closure of Coronary Artery Fistula

SAMUEL J. SHUBROOKS, JR., M.D., AND CHARLES Z. NAGGAR, M.D.

SUMMARY An asymptomatic 14-year-old male was found at cardiac catheterization to have a coronary artery fistula involving a vessel originating from the left main coronary artery and terminating in the right heart. Chest X-ray and electrocardiogram were within normal limits and shunt flow was too small to be detected by oximetry although a large vessel was seen angiographically. One year later, the previously loud continuous murmur had disappeared and repeat catheterization demonstrated near closure of the fistula. This is the first report documenting the spontaneous closure of a coronary artery fistula.

MANY CASES of coronary artery fistulae have been reported in the literature although this is a relatively uncommon entity. The majority of these reports describe the clinical and angiographic presentation and the immediate results of operative closure. However, little is known of the natural history of these fistulae and thus of the indications for and advisability of surgery. An important and previously unreported point in the spectrum of this anomaly is illustrated in the present case in which spontaneous near closure of a coronary artery fistula over a short time period was demonstrated.

Case Report

A 14-year-old male was first noted to have a heart murmur four years prior to admission. His medical history had

From the Departments of Medicine, New England Deaconess Hospital, The Lahey Clinic Foundation and Harvard Medical School, Boston, Massachusetts. Address for reprints: Samuel J. Shubrooks, Jr., M.D., 110 Francis Street, Boston, Massachusetts 02215. Received July 21, 1977; revision accepted August 18, 1977.
Cryoablation of drug-resistant ventricular tachycardia in a patient with a variant of scleroderma.

Circulation. 1978;57:190-197
doi: 10.1161/01.CIR.57.1.190

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/57/1/190

Permissions: Requests for permissions to reproduce figures, tables, or portions of articles originally published in Circulation can be obtained via RightsLink, a service of the Copyright Clearance Center, not the Editorial Office. Once the online version of the published article for which permission is being requested is located, click Request Permissions in the middle column of the Web page under Services. Further information about this process is available in the Permissions and Rights Question and Answer document.

Reprints: Information about reprints can be found online at:
http://www.lww.com/reprints

Subscriptions: Information about subscribing to Circulation is online at:
http://circ.ahajournals.org//subscriptions/