Atrial Fibrillation

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Atrial fibrillation is a supraventricular tachycardia characterized by a disorganized electrical activation and thus contraction of the atrium. The characteristic "irregularly irregular" rhythm is generated by the rapid irregular bombardment of the atrioventricular (AV) node with electrical impulses emanating from the atrial myocardium with variable conduction of the atrial impulses through the AV node (Figs. 9.1 and 9.2). The ventricular response is dependent on the ability of the AV node to transmit electrical signals (i.e., its refractory state). This is determined by its inherent electrophysiologic properties as well as autonomic tone.

Incidence

Atrial fibrillation is the most common rhythm disorder seen in man; prevalence in the general population is estimated at 0.4 %. There is a significant age-related distribution of the arrhythmia with 6 % of the population greater than 80 years of age suffering from atrial fibrillation. Atrial fibrillation remains an extremely rare arrhythmia in the pediatric population. Interestingly, genetic mutations in the KCNQ1 channel have been associated with atrial fibrillation as has the mutation related to the short QT syndrome (Figs. 9.3 and 9.4) has been reported (Chap. 18). Familial atrial fibrillation has involved an addition of nine different genetic defects from 13 loci; many of the genes are implicated in other cardiac arrhythmias (Chap. 19).

Associated Disease

Atrial fibrillation commonly occurs as a comorbid condition with other cardiovascular abnormalities. It is found in patients with congestive heart failure, mitral valve stenosis and insufficiency, hypertension, hyperthyroidism, and some forms of repaired congenital heart malformations. In addition, it is well recognized in the child and young adult with complex, functionally impaired congenital heart malformation, particularly those with extensive atrial incisions or suture lines (Figs. 9.5 and 9.6). Atrial fibrillation is also associated with Wolff–Parkinson–White syndrome. Atrial fibrillation occurring in conjunction with Wolff–Parkinson–White syndrome is a potentially life-threatening situation due to the lack of decremental conduction through the accessory pathway. Some accessory pathways are capable of rapidly conducting the atrial signals to the ventricle leading to ventricular fibrillation (Chaps. 4 and 21).

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M. Dick, II (ed.), *Clinical Cardiac Electrophysiology in the Young*,
DOI 10.1007/978-1-4939-2739-5_9

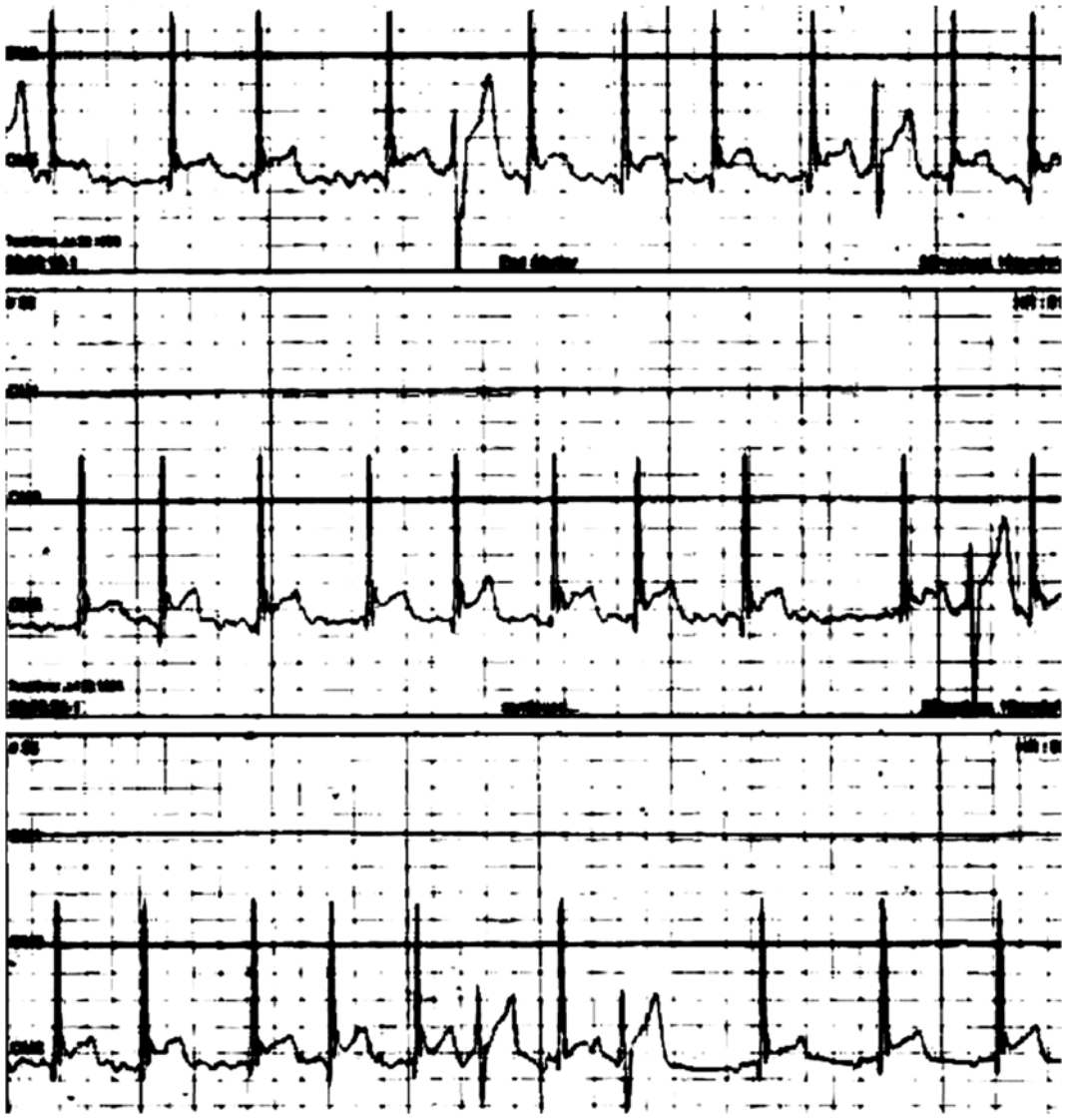

Fig. 9.1 Paroxysmal atrial fibrillation in a 17-year-old boy. Note the low amplitude fast waveforms (atrial fibrillatory waves) between the "irregularly irregular" QRS intervals. There are a few different QRS morphologies due to aberrant conduction. The atrial fibrillation spontaneously terminates (*bottom tracing*) as it frequently does in the paroxysmal form in patients without structural heart disease

Following elimination of the accessory pathway either surgically or via radiofrequency ablation, atrial fibrillation resolves in a large portion of these patients. Atrial fibrillation also may be associated with several other forms of supraventricular tachycardia in young patients. Our experience and that of others has been that young patients presenting with atrial fibrillation will frequently have other underlying mechanisms of SVT such as AVNRT, AVRT, ectopic atrial tachycardia, or atrial flutter. Ablation of these other tachycardia substrates results in the resolution of atrial fibrillation. Conversion of these forms of SVT with adenosine may result in transient atrial fibrillation (Fig. 9.7) as adenosine shortens atrial refractoriness.

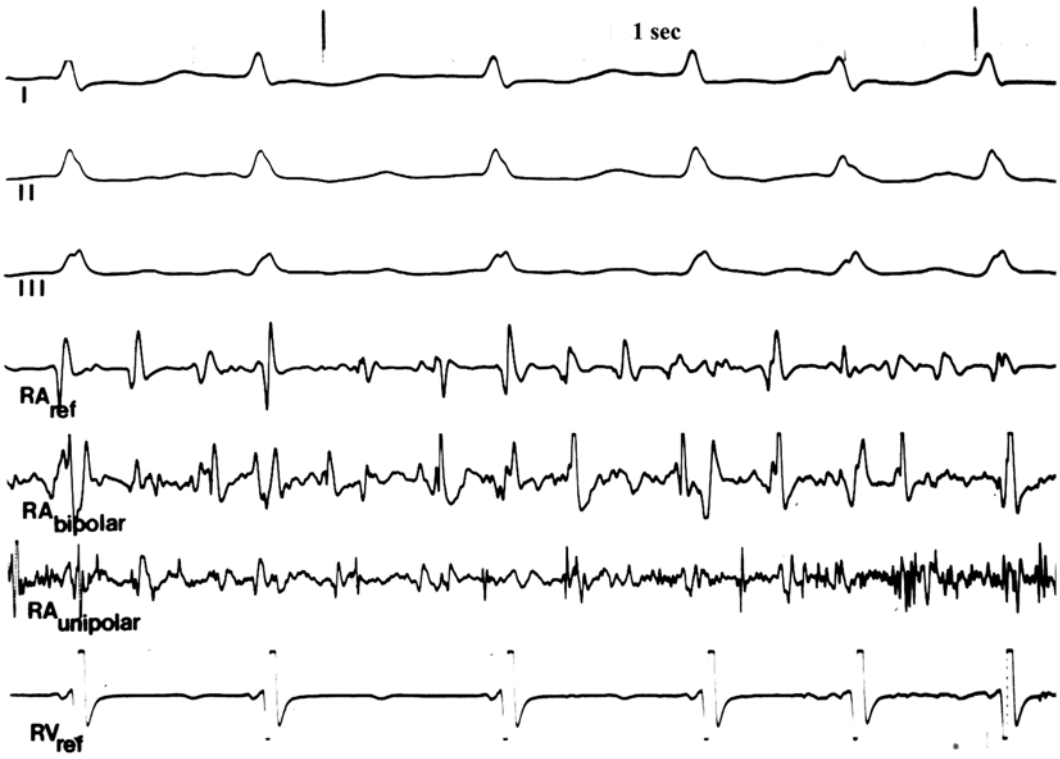

Fig. 9.2 Three electrocardiographic leads and four intracardiac electrograms [three in the right atrium (RA) and one from the right ventricular apex (RV)] illustrating the multi-phasic variable amplitude of the atrial fibrillation wave fronts

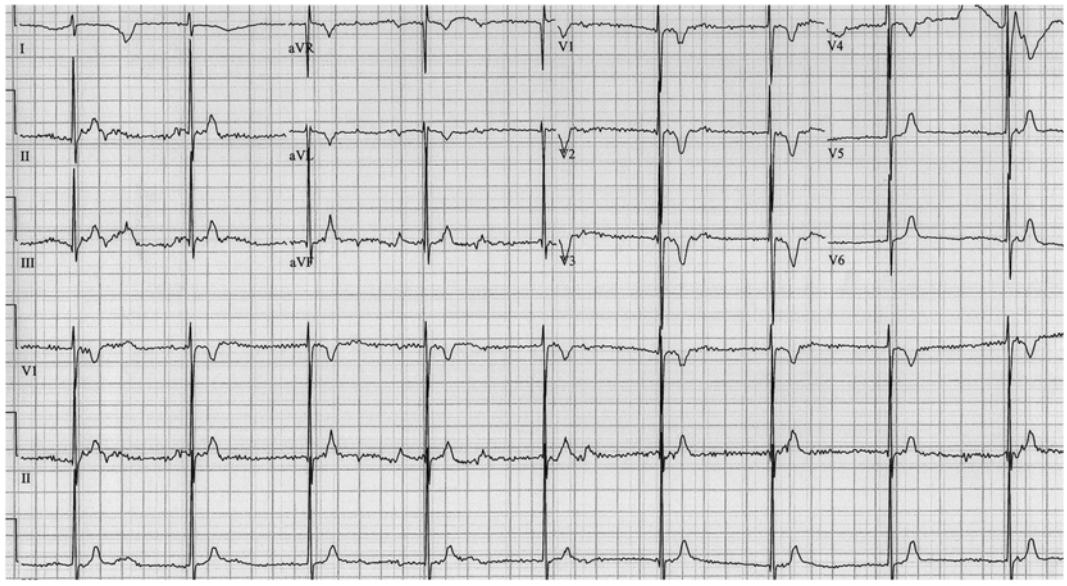

Fig. 9.3 One-week-old girl with congenital atrial fibrillation—the baseline shows a fine fibrillatory waveform, confirmed by esophageal electrogram (not shown)

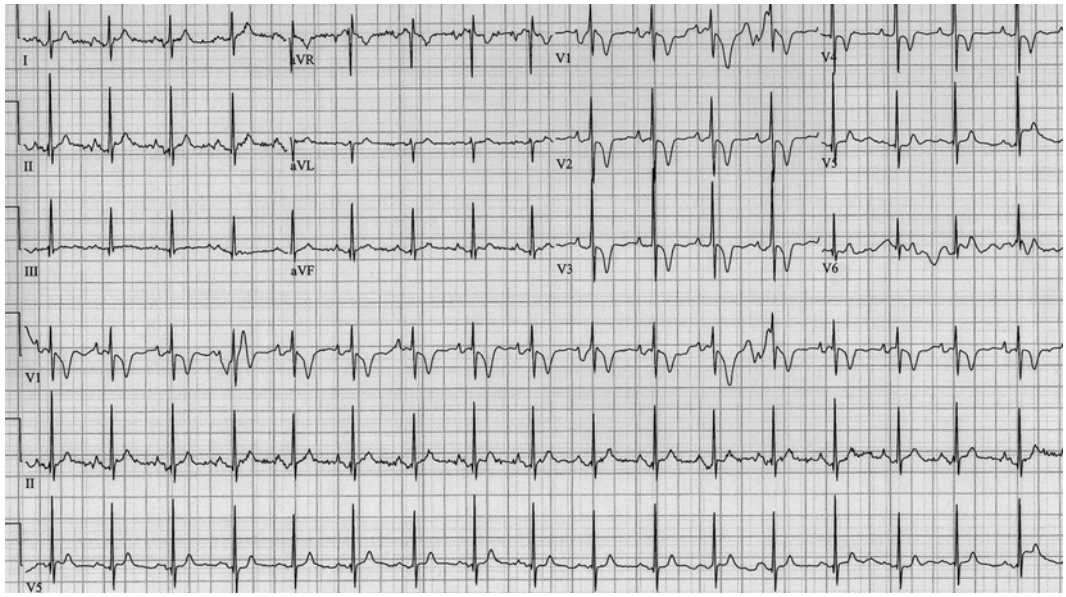

Fig. 9.4 Same patient as in Fig. 9.3—now in sinus rhythm (only for several hours; usual rhythm is atrial fibrillation). Note the very short QT interval (see text)

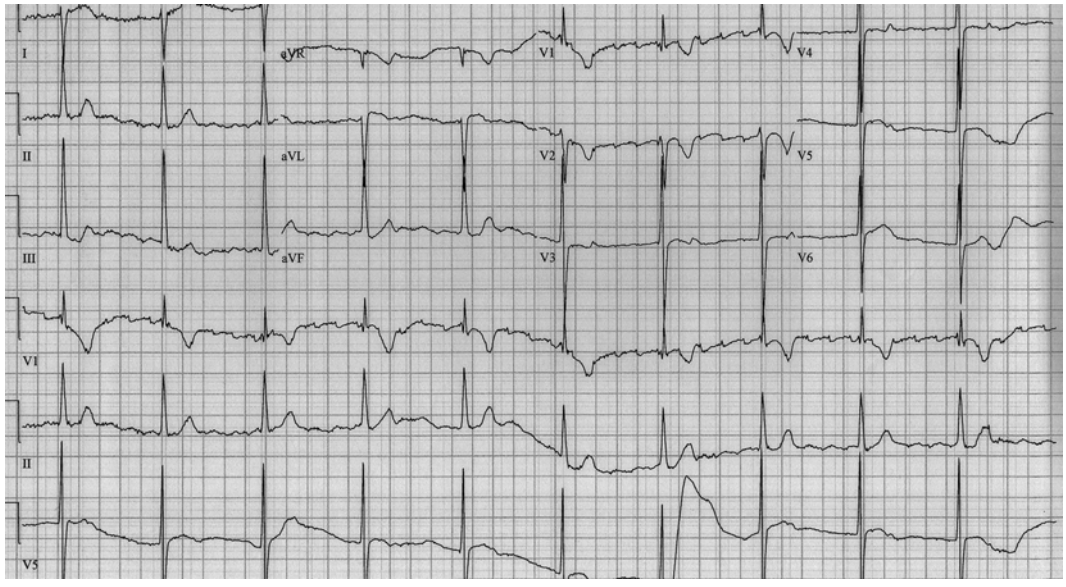

Fig. 9.5 Lead ECG in a 14 months boy after the double switch operation for congenitally corrected transposition of the great arteries. Note the atrial flutter-fibrillatory baseline

Another interesting association of atrial fibrillation is with obesity. This link has been demonstrated in adults as well as with pediatric patients. The exact physiological mechanism for this connection is unknown currently. We have also seen patients with atrial fibrillation that has probably resulted from enhanced vagal tone. Very strong vagal input can significantly shorten

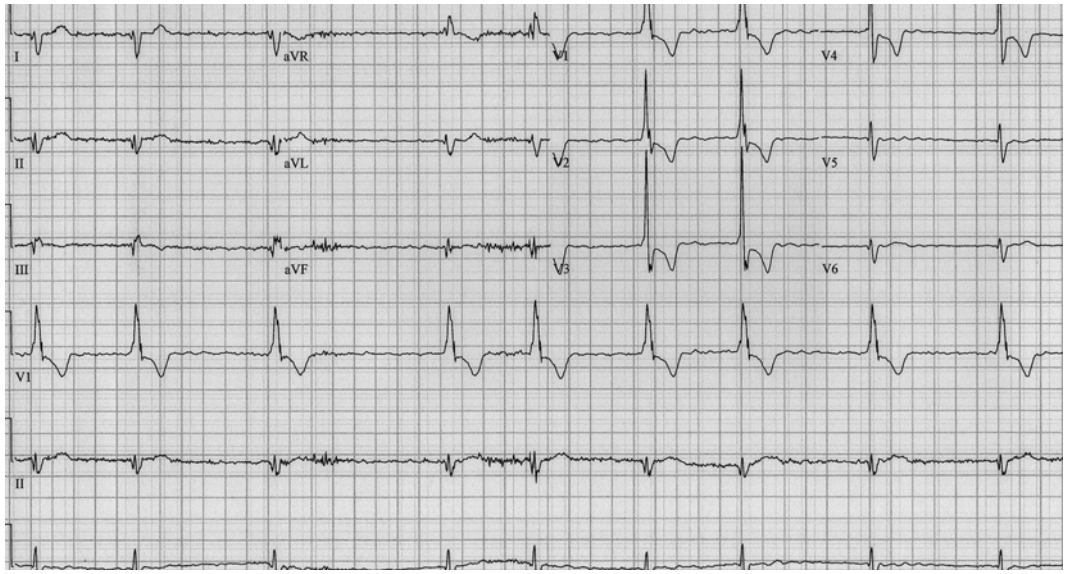

Fig. 9.6 12 Lead ECG from a 30-year-old man following the Mustard operation for D-transposition of the great arteries. Note the fine fibrillatory baseline and the "irregular irregularity" of the ventricular response

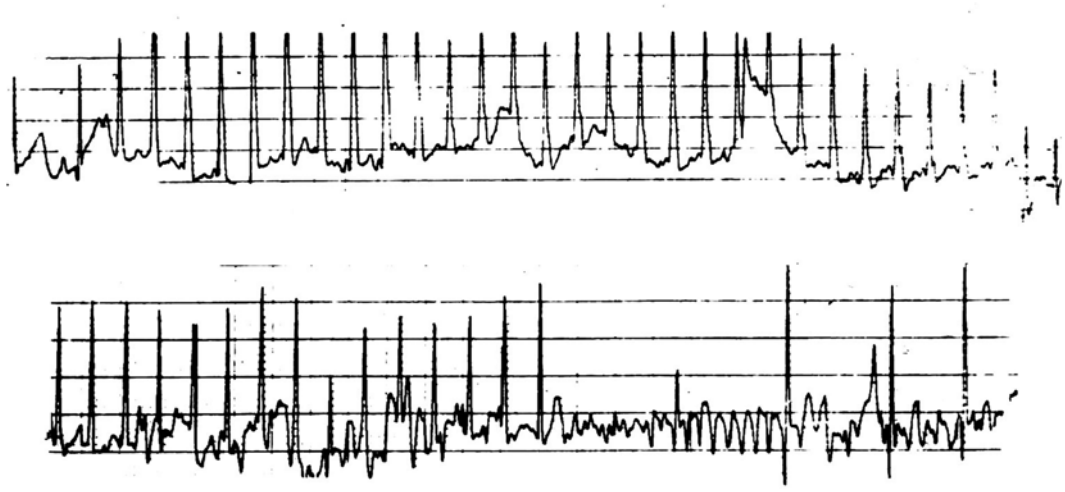

Fig. 9.7 Six hundred gram premature infant with SVT administered 120 mcg of adenosine. *Note*: The termination of the SVT but emergence of atrial fibrillation. The fibrillation spontaneously ceased several seconds later

the refractory period of atrial myocardium making the atria more susceptible to microreentrant circuits which underlie atrial fibrillation (see below). This is the physiological mechanism which leads to atrial fibrillation after adenosine administration (Fig. 9.7).

Recurrence

Two different recent reports demonstrated both a recurrence rate and an incidence of inducible supraventricular tachycardia in young patients

studied by programmed extra-stimulation of 39 %. Both of these findings support continued follow-up of these patients, if not electrophysiologic study after one episode of atrial fibrillation.

Mechanism

The pathophysiologic mechanisms underlying the initiation and perpetuation of atrial fibrillation remain under investigation (Fig. 9.2). It is possible that the mechanisms that support the initiation of atrial fibrillation are different from those that sustain the arrhythmia. The three most likely mechanisms include the multiple wavelet theory, single-circuit reentry, and multiple-circuit reentry with ectopic excitatory pulmonary vein foci. In this last model, multiple reentrant waves continuously circulate through the atrium using migrating central cores of refractory tissue to rotate around. The multiple wavelets circulate throughout the atrium following ever-changing pathways created by myocardium recovering from refractoriness. A critical mass of atrial tissue in this model is necessary to support a minimum number of wavelets in order to sustain atrial fibrillation. This has been cited as a rationale for the low incidence of atrial fibrillation in infants and children, as well as in small mammals (Chaps. 2 and 3). The second proposed mechanism for atrial fibrillation is the single-circuit reentrant model. In this model, a single "mother rotor" serves as the hub with multiple accessory circuits emanating from it. The third potential mechanism is atrial fibrillation arising from a rapidly discharging ectopic focus with fibrillatory conduction. This mechanism has come into favor with recent findings of excitatory loci occurring within the pulmonary veins, perhaps serving as the "trigger" of the "wavelets or of the "mother rotor." Recent clinical experience has demonstrated that focal radiofrequency ablation within the pulmonary veins as well as pulmonary vein isolation has successfully treated atrial fibrillation.

Sustained atrial fibrillation produces profound changes within the atrial myocardium, creating a substrate more conducive to supporting sustained atrial fibrillation. The electrical and structural remodeling of the atrium includes increasing fibrosis, as well as alterations in the expression of gap junctions and ion channels. These changes alter the mechanical and electrical properties of the atrial tissue by slowing the conduction velocity and a shortening of the refractory period in the atrium leading to tissue that is more likely to sustain atrial fibrillation.

The mechanism of the genetic mutation of atrial fibrillation and short QT appears to be a gain in function of the I_{Ks} in the KCNQ1 channel.

Therapy

Atrial fibrillation has proven to be an extremely difficult arrhythmia to manage with pharmacological therapy, achieving far less than optimal results. Multiple large clinical trials have been performed using nearly all of the currently available antiarrhythmic drugs, each of them with disappointing results. Large trials have also been performed comparing rate control (inhibition of conduction through the AV node) with rhythm control (restoration of sinus rhythm) for the ideal therapy. These studies have demonstrated no significant difference between these treatment options using total mortality, congestive heart failure, and rehospitalization as end points. Much of the difficulty with pharmacological therapy lies in the ever-present risk of ventricular proarrhythmia.

There are several non-pharmacological treatment strategies that have recently emerged. The Cox maze procedure creates multiple surgical incisions that are then repaired in the atrium in an attempt to channel the electrical signal between the sinus node and the AV-node while minimizing the formation of a reentrant loop. The surgical maze procedure has proven to have a reasonably high success rate, albeit with considerable associated surgical morbidity. An attempt to recreate this treatment principle using radiofrequency ablation has been attempted using long linear lesions, but with relatively poor results. The understanding of the role of rapidly discharging ectopic foci residing within the pulmonary veins has refocused the transcatheter therapies. Application of radiofrequency energy

or cryotherapy are now directed at either electrically isolating the pulmonary veins from the left atrium or at directly ablating the ectopic focus within the veins. A large meta-analysis published in 2013 sited a success rate of a single ablation procedure of just over 50 % for paroxysmal AF and just over 40 % for sustained AF. There was a considerable degree of variability in the reported results in the different studies cited. The long-term success rates (freedom from AF) after multiple procedures increased to almost 80 %, again with great variability between studies. When the total number of ablation procedures was evaluated, the average number of ablation procedures in the cohort was 1.5. A recent large study has demonstrated rhythm control (return to normal sinus rhythm) using radiofrequency ablation that is superior to either rate control or rhythm control using pharmacological agents. The difference between rhythm control using radiofrequency ablation and pharmacological agents likely rests in the proarrhythmia associated with pharmacological therapy.

Finally, pacemakers and implanted defibrillators have been used in an attempt to treat atrial fibrillation or to prevent its onset. To date, no significant improvement has been achieved relative to pharmacological therapies.

In conclusion, while atrial fibrillation is a common arrhythmia in older patients, it remains rare in pediatric and young adult patients. When encountered in pediatric patients, it often is the paroxysmal form with spontaneous remission and infrequent episodes. In addition, it can occur in the setting of other forms of supraventricular tachycardia, which are amenable to radiofrequency ablation therapy resulting in the resolution of the atrial fibrillation. For older patients with symptomatic or hemodynamic consequences, new ablation strategies aimed at eliminating or isolating the triggers within the pulmonary veins appears promising. Due to the infrequency of this arrhythmia in the young, transcatheter treatment targeted directly to atrial fibrillation is rarely indicated or necessary.

Suggested Reading

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