Focal atrial tachycardia (FAT) is an uncommon cause of supraventricular tachycardia in children, which occurs because of an abnormal, nonsinus, atrial focus with enhanced automaticity. 1–3 FAT often occurs as an incessant arrhythmia. The natural history includes possible progression to tachycardia-induced cardiomyopathy, which is reversible with control of the arrhythmia. 4–6

Clinical Perspective on p 670

Medical therapy has been the primary treatment for FAT, especially in young children who are more likely to have spontaneous resolution. 7,8 A variety of antiarrhythmic medications have been effective, although FAT is often resistant to pharmacological therapy. 7–12 Radiofrequency ablation (RFA) has been used successfully for definitive management. 13–16 Recently, the use of 3-dimensional electroanatomic mapping during catheter ablation has enabled improved rates of acute and long-term control. 13

The existing literature on FAT is limited to single-center experiences and small case series. There are no data to broadly evaluate the management and outcomes of FAT since the emergence and widespread use of modern antiarrhythmic medications and ablation techniques. This multicenter study describes the clinical course of a large, contemporary cohort of pediatric patients with FAT, including the prevalence of tachycardia-induced cardiomyopathy, effective medical management, and spontaneous resolution, as well as indications and outcomes for catheter ablation.

Methods

This is a retrospective review initiated by the coordinating site (Vancouver, Canada). Participating centers were solicited through the
Pediatric and Congenital Electrophysiology Society. Local ethics approval was obtained at each center.

**Patient Population**

Pediatric patients presenting with FAT between January 2000 and December 2010 were identified at 10 institutions. The diagnosis of FAT was based on ECG, 24-hour Holter, or event monitor data consistent with electrophysiological criteria previously described1,17: (1) narrow complex tachycardia with visible P waves at a rate inappropriate for age and activity, (2) identical abnormal P-wave morphology in the first and all subsequent tachycardia beats, (3) progressive increase in atrial rate with tachycardia onset (warm-up), (4) variable rate depending on autonomic tone, and (5) first- or second-degree atrioventricular block in the presence of continued tachycardia (Figure 1). Features that help distinguish FAT from sinus tachycardia include: (1) routine ECG atrial rate >150% of the predicted mean, (2) inverted and notched P wave in V1, (3) P-wave axis in the horizontal plane <0°, and (4) P-wave duration >90 ms in V1.18,19 Patients with significant structural heart disease were excluded.

**Data Collection**

Charts were reviewed for demographic characteristics, clinical presentation, occurrence of cardiomyopathy, medical and ablation therapy, and patient outcomes according to a standardized data collection form.

**Definitions**

FAT was considered incessant if demonstrated for >50% of monitored time by 24-hour Holter or telemetry. Cardiomyopathy was defined by an ejection fraction <40% or a shortening fraction <28%. Rhythm control was defined as normal sinus rhythm and noninducibility of the FAT after ablation. Resolution was defined as normal sinus rhythm on 24-hour Holter with no recurrence of symptoms after discontinuing antiarrhythmic therapy for 1 month. Spontaneous resolution was defined as resolution not requiring ablation therapy.

**Statistical Analysis**

Frequency tables were generated for all categorical data. Comparisons between groups of patients were made using a χ² test. Continuous data were analyzed using a univariate procedure. Data are presented as the median value with 95% distribution-free confidence limits constructed around the median value. A nonparametric 1-way ANOVA (Wilcoxon rank-sum test) was used to test for group differences. All statistical analyses were completed using SAS Statistical Software version 9.3 (SAS Institute, Cary, NC).

**Results**

**Patient Presentation**

The study population included 249 patients from 10 centers (Table 1). Median age at diagnosis was 7.2 (95% confidence interval [CI], 5.8–10.4) years with a median follow-up of 2.1 (95% CI, 1.8–2.6) years. There was a bimodal distribution of patients by age at diagnosis, with the highest number occurring from birth to 1 year (Figure 2).

FAT was detected as an incidental finding in the absence of other presenting symptoms or signs in 105 patients (42%). For children with symptoms, the most common presentations were palpitations (82 of 144; 57%), heart failure (24 of 144; 17%), and syncope (17 of 144; 12%). Fetal tachycardia was observed in 16 patients. Hospitalization was observed at the time of presentation for 101 patients (41%).

The diagnosis of FAT was based on ECG criteria in the vast majority of patients (99%). In 3 patients the diagnosis was based on Holter monitoring (n=2) or an event monitor (n=1). The median atrial and ventricular rates varied (atrial: 183 beats per minute; 95% CI, 170–197; ventricular: 176 beats per minute; 95% CI, 165–185) with atrioventricular block observed in 98 of 241 patients with tracings available for review (41%). P-wave axis was 0 to 90° in 56%. At presentation, FAT was incessant in 60 of 184 (33%). At diagnosis, echocardiography data were available for 210 patients (84%). The median shortening fraction was 34% (95% CI, 32–35%; n=202) and ejection fraction was 61% (95% CI, 59–63%; n=138).

**Initial Management**

Antiarrhythmic medications were used as initial therapy for 154 patients (62%; Figure 3). Catheter ablation was used for...
Spontaneous Resolution

Patients with spontaneous resolution were younger at presentation compared with patients who received catheter ablation (0.2 versus 12.2 years; \( P < 0.0001 \)). Fifty-three of 72 patients (74%) presenting at age <3 years had spontaneous resolution (Figure 2), including 50 patients aged <1 year at FAT diagnosis. Patients presenting at age <3 years were more likely to have spontaneous resolution compared with those presenting at age ≥3 years (74% versus 13%; \( P < 0.0001 \)). During follow-up, there were no cases of spontaneous resolution among 10 patients presenting from age 2 to 5 years. Spontaneous resolution was observed in 18 of 129 patients (14%) aged >5 years.
Fifty-one patients achieved resolution of FAT after a period of medical therapy. The median duration of therapy in these patients from initiation to discontinuation of all antiarrhythmic medications was 346 (95% CI, 307–428) days. The duration was similar for patients presenting at age <3 years compared with those presenting at age ≥3 years (337 versus 476 days; \( P=0.06 \)).

### Catheter Ablation

There were 134 patients (54%) who underwent a total of 167 catheter ablation procedures, including 69 patients who had ablation as initial therapy. The median age at first ablation was 12.8 (95% CI, 12.0–13.7) years. The median duration from diagnosis to first ablation was 62 (95% CI, 45–99) days. Effective catheter ablation was the most common indication, accounting for 55% of patients. Medical treatment failure or abnormal heart function occurred in the remaining. Seventy-three patients received ablation after medical therapy, including 31 patients with prior control of FAT using medication alone.

Ablation therapy was effective in 109 of 134 patients (81%; Table 3). Accounting for all 167 procedures, success and recurrence rates were 132 of 167 (79%) and 29 of 132 (22%). An electrophysiology study without catheter ablation was performed in 14 patients. Among these, 10 patients had FAT that was either suppressed or quiescent at the time of the procedure. Two patients with limited symptoms had ectopic foci in locations that were deemed to carry a significant risk of ablation (1 near the sinus node and the other near the atrioventricular node), so catheter ablation was not attempted. One patient had nonsustained FAT, and mapping was not sufficient for ablation to proceed. One patient experienced an air embolism during the electrophysiology study, and the procedure was stopped. Detailed procedural data were available for 148 procedures in 121 patients (90%). For these procedures, the ectopic focus was right-sided in 61%, including 25% in the right atrial appendage or crista terminalis. The remaining procedures showed foci in the left atrium (13%), left atrial appendage or crista terminalis (11%), pulmonary veins (9%), mitral valve annulus (1%), or other locations (5%). For left-sided foci, a transseptal approach was used in all procedures in patients as young as 2.8 years. Patient size was not a factor in determining a transseptal approach. For anesthetic management, 5 centers reported using general anesthetic only and 1 center reported using conscious sedation only. Four centers determined the anesthetic plan on a case-by-case basis, including starting with no/light sedation and progressing to general anesthesia as needed. The median procedure time was 193 (95% CI, 170–217) minutes, and the median fluoroscopy time was 23 (95% CI, 20–30) minutes with a median of 7 (95% CI, 6–9)
Table 3. Catheter Ablation for Focal Atrial Tachycardia

<table>
<thead>
<tr>
<th>Outcome</th>
<th>Ablation 1</th>
<th>Ablation 2</th>
<th>Ablation 3</th>
<th>Ablation 4</th>
</tr>
</thead>
<tbody>
<tr>
<td>Acute success</td>
<td>109 (82%)</td>
<td>17 (65%)</td>
<td>6 (75%)</td>
<td>0 (0%)</td>
</tr>
<tr>
<td>Partial success</td>
<td>5 (4%)</td>
<td>2 (8%)</td>
<td>0 (0%)</td>
<td>0 (0%)</td>
</tr>
<tr>
<td>Failure</td>
<td>18 (14%)</td>
<td>7 (27%)</td>
<td>2 (25%)</td>
<td>1 (100%)</td>
</tr>
<tr>
<td>Recurrence after</td>
<td>24 (22%)</td>
<td>3 (18%)</td>
<td>2 (33%)</td>
<td>...</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Indication</th>
<th></th>
<th></th>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Elective</td>
<td>74 (56%)</td>
<td>...</td>
<td>...</td>
<td>...</td>
</tr>
<tr>
<td>Treatment failure</td>
<td>49 (37%)</td>
<td>...</td>
<td>...</td>
<td>...</td>
</tr>
<tr>
<td>Abnormal heart function</td>
<td>30 (22%)</td>
<td>...</td>
<td>...</td>
<td>...</td>
</tr>
<tr>
<td>Other</td>
<td>10 (7%)</td>
<td>...</td>
<td>...</td>
<td>...</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Location</th>
<th></th>
<th></th>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>RA</td>
<td>84 (59%)</td>
<td>18 (60%)</td>
<td>4 (50%)</td>
<td>0 (0%)</td>
</tr>
<tr>
<td>LA</td>
<td>47 (33%)</td>
<td>9 (30%)</td>
<td>2 (25%)</td>
<td>0 (0%)</td>
</tr>
<tr>
<td>Multifocal</td>
<td>4 (3%)</td>
<td>1 (3%)</td>
<td>2 (25%)</td>
<td>1 (100%)</td>
</tr>
<tr>
<td>Noninducible/no data</td>
<td>7 (5%)</td>
<td>2 (7%)</td>
<td>0 (0%)</td>
<td>0 (0%)</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Mapping</th>
<th></th>
<th></th>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Electroanatomic mapping</td>
<td>98 (69%)</td>
<td>13 (43%)</td>
<td>5 (63%)</td>
<td>0 (0%)</td>
</tr>
<tr>
<td>Traditional mapping</td>
<td>44 (31%)</td>
<td>17 (57%)</td>
<td>3 (37%)</td>
<td>1 (100%)</td>
</tr>
</tbody>
</table>

*Two patients with EPS only, no ablation during the first catheter procedure went on to have catheter ablation during a subsequent procedure, accounting for 134 patients who underwent catheter ablation overall.

lesions ablated for a median total RFA time of 287 (95% CI, 221–360) seconds. Patients were followed for a median of 21.7 (95% CI, 17.0–29.0) months after ablation.

Three-dimensional electroanatomic mapping was used in 98 of 134 patients (73%) who underwent ablation, accounting for 110 of 167 of all procedures (66%). Similar rates of acute success were achieved for patients who underwent ablation with electroanatomic and conventional mapping (81% versus 75%; P=0.42); however, recurrence after acute success was less likely in the electroanatomic mapping group (16% versus 35%; P=0.02). Electroanatomic mapping was also associated with similar rates of acute success (84% versus 79%; P=0.62) and lower rates of recurrence (14% versus 42%; P=0.004) for all first-time catheter procedures. Patients undergoing ablation with electroanatomic mapping were more likely to ultimately achieve FAT control when compared with patients undergoing ablation with conventional mapping alone (96% versus 74%; P<0.01).

Thirty patients underwent multiple catheter procedures because of FAT recurrence (n=16), initial ablation failure (n=12), or inability to induce FAT at the initial electrophysiological study (n=2). These 30 patients had 35 repeat ablation procedures. Compared with initial RFA, repeat ablation was less effective (83% versus 66%; P=0.037). Recurrence rates were similar (22% versus 22%; P=0.77).

Complications of RFA were observed in 8 patients. A 15-year-old boy experienced cardiac arrest 6 hours after ablation, which was attributed to bradycardia-related torsades de pointes. Another 15-year-old boy experienced air embolism during his electrophysiology study, and the procedure was stopped. A third 15-year-old boy experienced a stroke during his second ablation procedure. A 2-month-old boy developed sinus node dysfunction after ablation, and a 7-year-old boy experienced pulmonary vein stenosis after RFA of an ectopic focus at that location. Three patients experienced other arrhythmias in the context of RFA procedures, including 2 with atrial fibrillation and 1 with atrial flutter, all requiring electric cardioversion.

**Cardiomyopathy**

Echocardiographic evidence of tachycardia-induced cardiomyopathy was observed in 69 patients (28%). The median shortening fraction and ejection fraction in these patients were 21% (95% CI, 17–23%; n=63) and 40% (95% CI, 32–44%; n=41), respectively. Normal heart function was achieved in 58 of 67 patients (87%) >3.3 months (95% CI, 1.1–4.5 months; n=47). Two patients did not have a repeat echocardiogram. Clinical improvement was secondary to rhythm control in 46 of 53 patients (87%). Medications to improve heart function and cardiomyopathy-related symptoms including intravenous inotropic agents, diuretics, and angiotensin-converting enzyme inhibitors were used in 36 of 69 patients (52%). Three patients (1-day-old girl, 3-month-old boy, 13-year-old girl) required extracorporeal life support in the context of severe cardiac dysfunction with FAT refractory to treatment. One of these patients (1-day-old girl) ultimately underwent cardiac transplantation.

Fifty-five of 69 patients (80%) with cardiomyopathy had resolution of FAT at last follow-up. Eleven additional patients continued on antiarrhythmic therapy with suppression of FAT. Resolution was similar in patients with and without evidence of cardiomyopathy (95% versus 87%; P=0.13).

**Patient Status**

At last follow-up, 185 of 209 patients (89%) had resolution of FAT. An additional 40 patients continued to receive antiarrhythmic medications with no evidence of arrhythmia. In 71 of 209 patients (34%), resolution was observed without catheter ablation.

There were 9 patients with persistent FAT despite ongoing medical treatment. One patient received concomitant therapy for cardiomyopathy. Two patients continue to be treated for cardiomyopathy alone, and 14 continue with FAT but with no medical treatment. As mentioned above, 1 of the patients on extra corporeal life support (a 13-year-old girl) died after support was withdrawn because of significant neurological complications.

**Discussion**

This multicenter study describes the clinical course and outcomes for 249 children with FAT. To our knowledge, this is the largest study of FAT in children and reflects the scope of current therapeutic options.

For this patient cohort, FAT management is characterized by an increased number of therapeutic options compared with previous data, including the number and combinations of antiarrhythmic medications, as well as the availability and use...
of catheter ablation therapies.2,3,7–11,14,20 Patients in this study received 44 different medication combinations. In total, 54% of all patients received at least 1 RFA procedure, including 4 procedures in children aged <1 year. Overall, the rate of FAT resolution was 89%.

As a result of these expanded options, the calculus for clinical decisions has changed. Patients presenting at any age may receive primary medical or ablation therapy followed by, if necessary, subsequent trials of either therapeutic modality. In some cases, children with FAT refractory to initial treatments undergo a complex clinical course involving multiple medications and catheter procedures. With emerging experience necessary, subsequent trials of either therapeutic modality. In

Our study shows that 74% of children with FAT diagnosed in the first year of life achieve spontaneous resolution, suggesting that these children, in particular, may benefit from an initial trial of medical therapy. The rate of spontaneous resolution reported in this group is a conservative estimate. For some children, primary or early ablation therapy interrupts the possibility of spontaneous resolution and, therefore, alters the natural history. Among older children, the rate of spontaneous resolution is more difficult to assess reliably because of the increased use of catheter ablation in this group. These findings are similar to previous reports that showed spontaneous resolution in a majority of patients, with higher rates among young children.7,8,11,20

In terms of medical therapy for FAT, there are no clear trends to suggest which medications are most useful. Overall, 22 different medication combinations were effective for FAT suppression. Current medication choices depend on physician preference and empirical trials. In some cases, FAT may be difficult to treat and require multiple medication combinations; however, >70% of patients who receive medical therapy eventually achieve control. Some patients have ongoing FAT when medication is withdrawn and require subsequent medical or catheter therapy. The option for a sustained period of FAT control with medical therapy has an important role in preventing or reversing the functional changes associated with tachycardia-induced cardiomyopathy.

Our study suggested that β-blockers and class Ic antiarrhythmic medications were most commonly useful for FAT management compared with other medications such as amiodarone that were less useful. However, our analysis includes relatively few cases for comparison and, as a retrospective study, has no standard dosing or criteria for medication failure or changing medication regimens. A prospective, controlled trial to determine which medications are most effective and in which patient groups may be needed.

Although spontaneous resolution in older children is uncommon, this study confirms that RFA is useful for this group. Many older patients in our study received successful primary ablation therapy. Overall, RFA was effective as definitive therapy in 80% of all FAT patients. Catheter ablation has previously been shown to be a safe and effective therapeutic option for pediatric supraventricular tachycardia, including FAT.15,24 Our success and recurrence rates are comparable to those reported in the literature.5,11

There is limited experience with catheter ablation of FAT in younger children. Although initial studies of RFA in young or small children suggested there may be lower success and higher complication rates,15,22,23 more recently, Blaufox et al25 found similar success and complication rates for catheter ablation of FAT in children aged <1.5 years compared with older children. In our study, the youngest patient to receive ablation therapy was aged 2 months. Catheter ablation was successful for FAT resolution after unsuccessful medical therapy in the setting of tachycardia-induced cardiomyopathy.

Our study also supports evidence that electroanatomic mapping instead of conventional mapping for catheter ablation improves outcomes in pediatric FAT. Toyohara et al21 reported a series of 35 pediatric FAT patients who had 100% acute success and 11% recurrence with RFA using the CARTO Navigation System (Biosense Webster, Inc, Diamond Bar, CA). Data from Cummings et al13 showed improved ablation success and recurrence rates with electroanatomic mapping compared with conventional mapping for pediatric FAT. Predictably, electroanatomic mapping techniques are associated with reduced fluoroscopy time26,27 and have been suggested to improve catheter ablation of pediatric arrhythmias, particularly for complex substrates including patients with structural heart disease.28

Study Limitations

This is a retrospective study and subject to limitations, including variable duration of follow-up and available data for each patient, as well as a bias toward selecting challenging cases and cases managed with ablation at tertiary care centers. As the largest series of FAT to date, this study represents a broad spectrum of patients and practice patterns. Specific information regarding medication dose and procedural details for each ablation procedure was not assessed.

Conclusions

FAT is managed successfully in most children despite no standardized approach. Children with FAT often present with incessant tachycardia and tachycardia-induced cardiomyopathy. Spontaneous resolution is common, especially in young children, and these children are likely to benefit from an initial trial of medical therapy to reduce arrhythmia burden. Catheter ablation is frequently successful with fewer recurrences when using electroanatomic mapping techniques.

Sources of Funding

This work was funded by a grant from the Rare Disease Foundation, Vancouver, Canada.

Disclosures

None.
References


CLINICAL PERSPECTIVE

Focal atrial tachycardia in children often presents in infancy and may resolve or persist and even progress to tachycardia-induced cardiomyopathy. Data on therapy and outcomes are limited. In this large, contemporary, retrospective observational series, medical management was the mainstay of treatment in younger patients, and combination therapy was prescribed in nearly half of the patients. No single regimen appeared most effective. Catheter ablation was used as initial therapy in 28% of patients. Resolution of tachycardia-induced ventricular dysfunction was achieved in nearly all patients (87%). The overall outcome was good for this patient population with few adverse events. Although outcomes are favorable, studies are needed to clarify optimal treatment approaches for young patients with this arrhythmia.
Current Management of Focal Atrial Tachycardia in Children: A Multicenter Experience

Circ Arrhythm Electrophysiol. 2014;7:664-670; originally published online July 11, 2014; doi: 10.1161/CIRCEP.113.001423

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circep.ahajournals.org/content/7/4/664

Permissions: Requests for permissions to reproduce figures, tables, or portions of articles originally published in Circulation: Arrhythmia and Electrophysiology 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: Arrhythmia and Electrophysiology is online at:
http://circep.ahajournals.org//subscriptions/