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Review

# Atrial flutter and fibrillation in the young patient without congenital heart disease ☆

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ABSTRACT

Atrial flutter and fibrillation are infrequently encountered in the young patient without congenital heart disease. Lone atrial flutter appears to be more common in boys and is most often present at birth. Cases diagnosed beyond the first week of life may present with tachycardia-induced cardiomyopathy, which typically resolves upon restoration of sinus rhythm. While antiarrhythmic drug therapy and catheter ablation may be indicated in some, most patients experience no recurrence on follow-up. Lone atrial fibrillation, though equally rare in children, typically presents in adolescence. In the absence of structural heart disease, atrial fibrillation in the young most frequently occurs in patients with Wolff–Parkinson–White syndrome or channelopathies, such as long and short QT and Brugada syndromes. Atrial fibrillation may also be triggered by acquired or iatrogenic conditions that result in atrial remodelling, inflammation, infiltration, and/or autonomic or neuro-hormonal imbalances. In the absence of pediatric evidence-based guidelines, management decisions regarding antiarrhythmic and anticoagulation therapy are largely inferred and adapted from the adult literature. Many questions remain unanswered, including thromboembolic risk estimates, pathophysiological mechanisms, potential associations with environmental factors such as competitive sports, and underlying genetic determinants.



## 1. Introduction

Although prevalence estimates remain imprecise, atrial flutter and fibrillation are uncommon rhythm disorders in the young patient without congenital heart disease. For example, in a multicenter study that included 380 patients with atrial flutter between 1 and 25 years of age, only 8% had structurally normal hearts [1]. The lack of large pediatric studies addressing these arrhythmias precludes evidence-based recommendations. Associated morbidity, including thromboembolic risk, is poorly defined such that clinical decisions regarding anticoagulation therapy are largely based on extrapolations from adult studies. Within this context, this review summarizes the current state of knowledge regarding the scope of the problem, electrocardiographic findings, clinical presentations, management options, and long-term prognosis. To maximize clinical relevance, practical issues are emphasized. Atrial flutter is discussed separately from atrial fibrillation and special circumstances are addressed, including channelopathies, genetic factors, and Wolff–Parkinson–White (WPW) syndrome.

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## 2. Atrial flutter

### 2.1. Lone atrial flutter in newborns and infants without congenital heart disease

#### 2.1.1. Epidemiological features

Lone atrial flutter is rare in newborns and infants. As illustrated in Fig. 1, improvements in fetal echocardiography have permitted prenatal diagnoses in some. In fact, atrial flutter now accounts for roughly 25% of detected fetal tachyarrhythmias [2–4]. Since the 1950s, several case series have provided insights into epidemiological features and natural history. The largest series report a male predominance, with a 2:1 to 3:1 ratio of boys to girls [5–7]. Some authors have distinguished congenital atrial flutter, i.e., diagnosis prenatally or during the first week of life, from those recognized beyond the first week of life [6,8]. The majority of reported cases are congenital, with 75% of all patients diagnosed before 2 days of age [5,7].

#### 2.1.2. Electrocardiogram findings

In most patients, the diagnosis is clinched by an electrocardiogram (ECG) that demonstrates classic flutter waves. In the case of uncertainty, transesophageal electrocardiographic recordings may provide valuable diagnostic information [9]. Atrial rates typically vary between 300 and 500 beats per minute (bpm), with the fastest rates in the fetus. One-to-one atrioventricular (AV) conduction is

uncommon, as over 75% of cases have a 2:1 AV relationship [4,7,9]. Consequently, ventricular rates vary between 150 and 250 bpm.

When atrial flutter coexists with other arrhythmias, risk of recurrence appears higher [7]. Whereas concomitant atrial fibrillation and ectopic atrial tachycardia are infrequent, accessory pathway-mediated supraventricular arrhythmias have been reported in 14% to 33% of cases [7,8,10,11]. It remains uncertain whether they share common predisposing and/or etiological factors, or whether accessory pathway-mediated arrhythmias or conduction beget atrial flutter [7,11]. Some authors have speculated that atrial flutter requires a critical atrial mass of tissue to propagate and that prolonged periods of supraventricular tachycardia may result in structural and electrical remodelling that favor atrial macroreentry. Others have hypothesized that ventricular premature beats that are conducted retrogradely and eccentrically through an accessory pathway may facilitate initiation of atrial flutter [11].

*2.1.3. Clinical presentation*

The clinical presentation appears to be "bimodal", i.e., either as an incidental finding in asymptomatic patients (80%) or in the setting of moderate to severe heart failure (20%) [7]. In asymptomatic patients, tachycardia is typically detected by routine examination and monitoring during the first few hours or days of life [7]. Episodes are usually brief and conversion to sinus rhythm may be obtained without difficulty.

The advanced heart failure group includes those with refractory fetal atrial flutter and hydrops. Prompt delivery by either vaginal induction or Cesarean section is usually indicated. In a study of 44 fetuses with a prenatal diagnosis of atrial flutter at a median gestational age of 31.5 weeks (range 19–40 weeks), fetal hydrops was seen in 20 (45%) cases [12]. The associated mortality and neurological morbidity was 9% each. In contrast, no death occurred in non-hydropic fetuses, underscoring the importance of this prognostic marker. Beyond the newborn period, most cases of atrial flutter are diagnosed in infants with tachycardia-induced cardiomyopathy [9]. The clinical history typically reveals feeding difficulties and tachypnea for days or weeks prior to presentation. Ventricular systolic dysfunction and atrial dilation may be readily appreciated by echocardiography. With restoration of the sinus rhythm, ventricular dysfunction resolves within a matter of weeks in nearly all patients [7].

*2.1.4. Treatment and long-term prognosis*

Management and therapeutic options have evolved over the past 30 years. In the 1970s, pharmacological therapy (particularly digoxin) was considered the first line treatment for acute episodes of atrial flutter [6,8,13]. Today, a rhythm-control strategy with electrical cardioversion or transesophageal overdrive pacing is generally favored [7,9,11,14]. In asymptomatic infants with normal ventricular function, delayed cardioversion (e.g., up to 24 h) may be reasonable given that 25% convert spontaneously [7]. Whereas long-term management typically consisted of digoxin therapy for a few months to a year or more, it is now known that atrial flutter most often occurs as a single episode with no late recurrence. As such, long-term prophylaxis is no longer routinely recommended [5,7,14]. For patients with recurrent episodes, particularly older children, it may be reasonable to consider flutter ablation as an alternative to drugs given the low procedural risks, high efficacy, and often-unsatisfactory response to pharmacological agents [15]. Three-dimensional electroanatomic maps of atrial flutter are shown in Fig. 2, with catheter ablation illustrated in Fig. 3.

## 3. Atrial fibrillation

*3.1. Lone atrial fibrillation*

Atrial fibrillation is the most common sustained arrhythmia in adults, with an overall prevalence of 1% that continues to increase with age [16]. Approximately 10–30% of subjects have no apparent

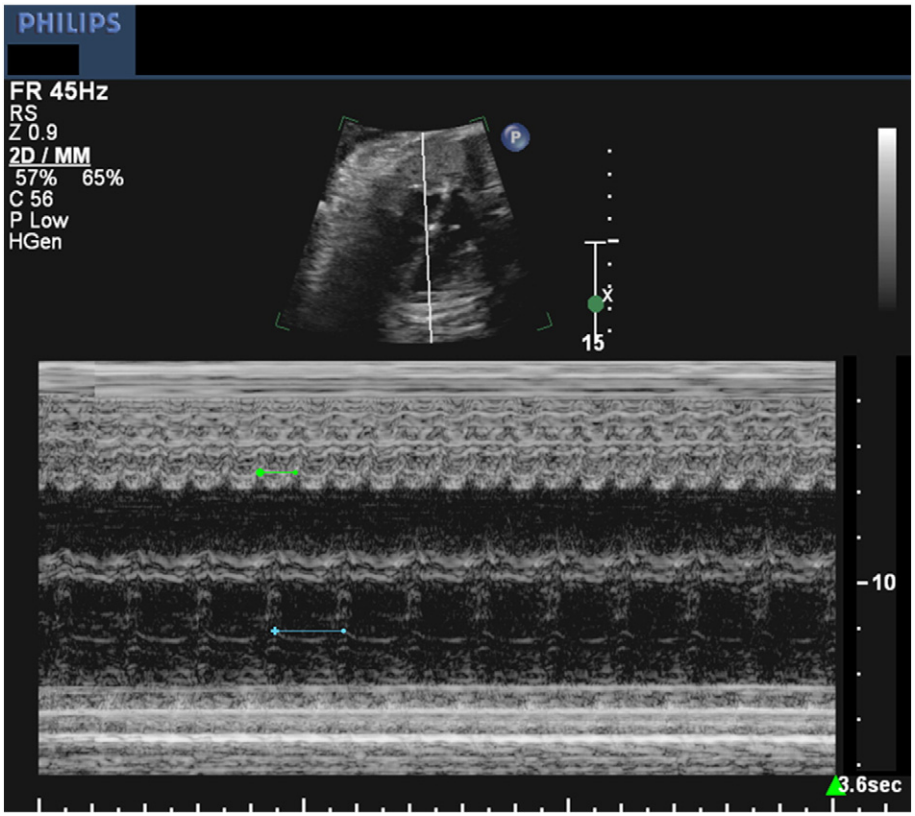


**Fig. 1.** M-mode (right atrium/left ventricle) fetal echocardiogram performed at 38 weeks of gestation demonstrating atrial flutter with a cycle length of 130 ms (green interval above) and a 2:1 atrioventricular response (blue interval below). The atrial flutter converted spontaneously during induced vaginal delivery, with no recurrent episode.