

## CASE STUDY

## Familial neonatal atrial tachycardia

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**Abstract**

**A father and his two sons each presented with atrial tachycardia in the newborn period. The father went on to develop dilated cardiomyopathy. The first son (who also had transposition of the great arteries) died from the arrhythmia after surgery. The second son is currently successfully managed pharmacologically.**

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Atrial tachycardia is a rare but serious arrhythmia. It is often unresponsive to pharmacological treatment and to direct current cardioversion.<sup>1</sup> The arrhythmia tends to be incessant<sup>1</sup> and can lead to cardiac dilatation and failure.<sup>2</sup> It is also rare for the arrhythmia to present in the neonatal period.<sup>3</sup> We report a family of a father and his two sons, each of whom had an incessant atrial tachycardia presenting in the neonatal period.

**Patients**

The father, a member of the Irish travelling community, presented in 1971 at the age of three weeks with cardiac failure and incessant tachycardia. He was the last of five children, and there was no similar history in his parents or his siblings. The ventricular rate was 200 beats/min and the arrhythmia was diagnosed

as atrial flutter. Cardioversion was unsuccessful. He was put on digoxin for ventricular rate control and was followed up. Incessant tachycardia persisted with a variable ventricular response. The poor quality of the available electrocardiograms did not permit an accurate diagnosis to be made. However, there was evidence of a narrow complex tachycardia with lack of a 1:1 P to QRS relation (fig 1). The failure of electrical cardioversion suggests in retrospect that the tachycardia mechanism was one of abnormal automaticity rather than reentry. Subsequent electrocardiograms recorded since 1985 showed an isoelectric baseline suggesting atrial fibrillation. Although he has remained well, chest radiographs and echocardiograms have shown cardiac dilatation since he was six months old. Currently he is in atrial fibrillation with an acceptable ventricular rate. Cross sectional echocardiography, however, shows a dilated, poorly contractile left ventricle. He is being treated with anticoagulants but does not require anti-failure therapy. He is aware that he may need a cardiac transplant in the future.

His first son, born in 1989, presented with cyanosis at the age of two days and was found to have transposition of the great arteries. After balloon atrial septostomy, an incessant narrow QRS complex tachycardia of 200 beats/min developed. The heart rate was unvarying, and no atrioventricular block was noted. With intravenous verapamil a 2:1 atrioventricular block was produced (fig 2). The initial negativity of the P waves in lead I suggested a left atrial origin. The diagnosis was confirmed at electrophysiological study and

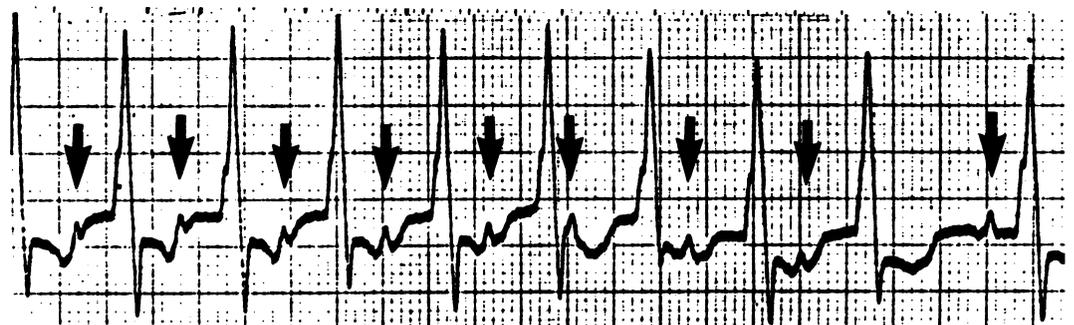


Figure 1 Electrocardiogram recording (50 mm/s paper speed) of lead V1 in the father showing positive P waves (arrows). Note the prolonged and variable PR interval, with 1:1 conduction maintained most of the time.

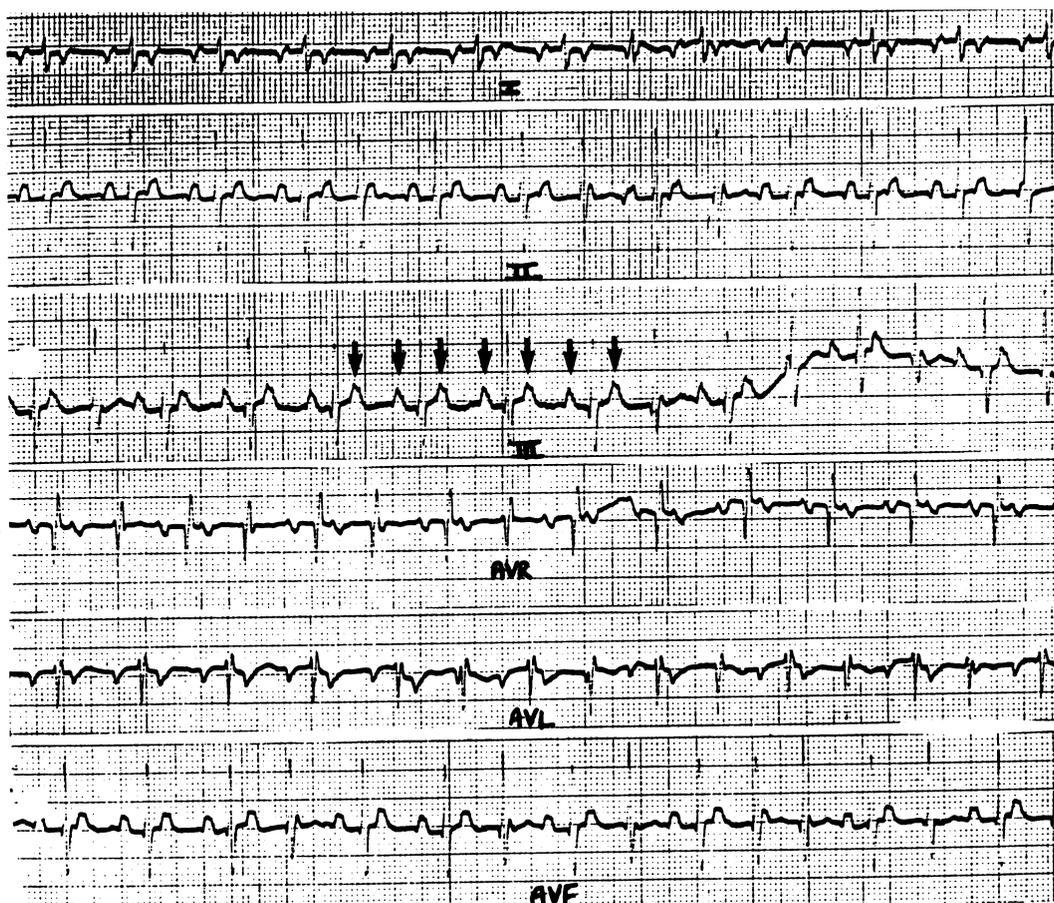
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Figure 2  
Electrocardiogram recording (25 mm/s paper speed) of the frontal leads in the first son. Note the 2:1 relation between P waves (arrows) and QRS complexes.



the rhythm was found to originate high in the lateral right atrium. However, an origin in the left atrium around the orifice of the right upper pulmonary vein could not be excluded.

The arrhythmia could not be controlled pharmacologically despite various combinations of digoxin, propranolol, flecainide, and amiodarone. Surgical cryoablation (without map-

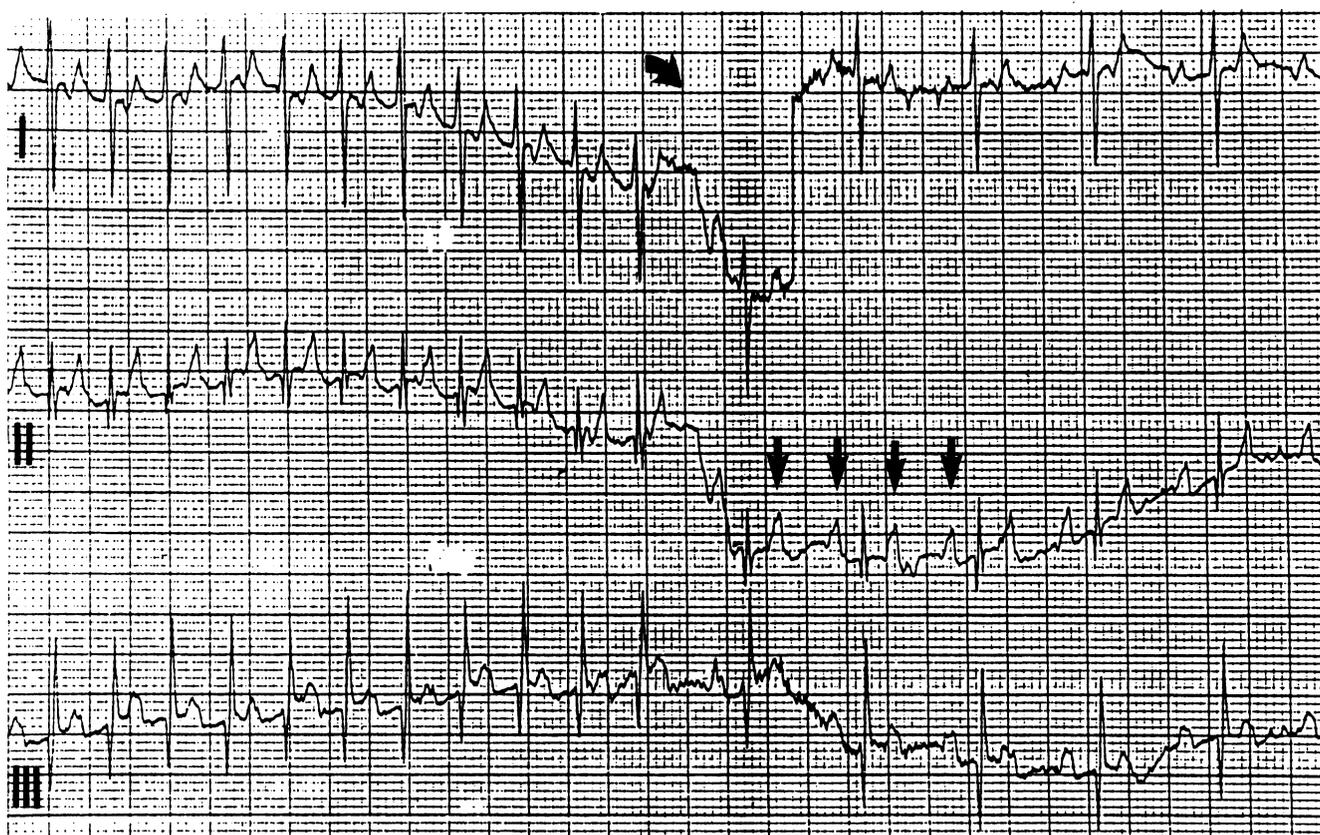


Figure 3 Electrocardiographic recording (25 mm/s paper speed) showing development of 2:1 atrioventricular conduction with adenosine injection. Note two P waves (small arrows) to each QRS complex after the large arrow.

ping) was attempted but failed. After this operation he deteriorated significantly. Moderate surface hypothermia was used to achieve rate control and haemodynamic stability. This aspect of the case has already been reported in a paper on the use of hypothermia in children with intractable arrhythmias.<sup>4</sup> At the age of six weeks, a Senning operation was performed, with the child still in incessant atrial tachycardia, but he died in the operating theatre.

The second son, born in 1993, had prenatal tachycardia at a rate of 240 beats/min. Although he was in sinus rhythm at birth, a tachycardia of 275 beats/min was noted the next day. With adenosine injection a 2:1 block occurred (fig 3). The P wave morphology was similar to that found in his brother (figs 2 and 3) with an initial negative deflection followed by a positive P wave in lead I. Treatment with flecainide was started and subsequently propranolol was added, with successful conversion to sinus rhythm. At follow up he was in sinus rhythm on a standard 12 lead electrocardiogram and 24 hour monitoring electrocardiogram, with normal cardiac structure and function on cross sectional echocardiography.

### Discussion

Atrial tachycardia is a rare arrhythmia. It is thought to be caused by abnormal automaticity. Some use the term automatic atrial tachycardia. Because the tachycardia originates from a focus other than the normal sinus node, another term used for this arrhythmia is ectopic atrial tachycardia.<sup>5</sup>

All the three patients in our report had a tachycardia confined to the atrium. The tachycardia was not caused by reentry in any of the three because it showed no response to DC cardioversion in two and to atrioventricular node blockers in two. The two sons also had similar and abnormal P waves, suggesting an ectopic origin for the tachycardia. Therefore, the tachycardia was probably caused by abnormal automaticity and also an ectopic focus. However, we have chosen to use the broader term "atrial tachycardia" because the exact nature was not established.

The first child seemingly developed the arrhythmia after a balloon atrial septostomy. Mechanical injury may have helped to cause the arrhythmia, but this explanation seems unlikely. Firstly, at electrophysiological study the tachycardia was not found to originate in the septum, and secondly this is not a com-

monly reported arrhythmia after atrial septostomy. It may be that this baby had an underlying abnormality that was triggered by the procedure.

Although radiofrequency catheter ablation can achieve a permanent cure<sup>6</sup> for this condition, we elected to continue drug treatment in the last child. This approach was preferred because of the higher risks of catheter ablation in small babies, and also because the arrhythmia can resolve spontaneously if treatment is successful for more than one year.<sup>7</sup>

Atrial tachycardia is a rare arrhythmia. However, its recognition is important because it can be incessant and result in cardiomyopathy. The exact incidence of the arrhythmia is unknown. In the Pediatric Radiofrequency Association Registry,<sup>6</sup> 38 (5.8%) of 652 patients had atrial tachycardia.

Familial forms of both bradyarrhythmias and tachyarrhythmias have been described,<sup>8</sup> including familial atrial fibrillation<sup>8</sup> and atrial flutter.<sup>9</sup> Atrial arrhythmias have been associated with familial arrhythmogenic right ventricular dysplasia<sup>10</sup> and with familial atrioventricular block.<sup>11</sup> However, we were unable to find a previous report of familial atrial tachycardia.

In summary, we believe this is the first report of what is possibly a familial form of atrial tachycardia presenting in the neonatal period.

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