

# Establishing a pathway for the diagnosis and management of arrhythmias in patients with pulmonary hypertension

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Pulmonary hypertension (PH) is a devastating, life-limiting disease driven by small vessel vascular remodeling and can lead to right heart failure and death.<sup>1</sup> Pre-syncope, syncope and palpitations are frequently described by patients, however a firm diagnosis of arrhythmia is often not reached using 12-lead ECG and Holter monitoring. Holter monitors are often requested to be performed at local hospitals for patient ease but the test is often not performed or results are not fed back to the PH team. Insertable cardiac monitors (ICM) permit remote monitoring of a patient's heart rate, heart rate variability, heart rhythm and physical activity. When an arrhythmia is identified we know that guideline-based treatment of arrhythmias improves quality-of-life and prognosis in left heart disease but these guidelines are not PH-specific and this can make arrhythmic management in this cohort complicated.

## OBJECTIVES

1. Set up a clinical ICM service at Sheffield PVDU to improve access to remote continuous monitoring of arrhythmias in patients with PH and increase diagnostic accuracy in symptomatic patients.
2. Use knowledge obtained to determine a treatment pathway for arrhythmias that is specific to and safe the PAH population.

## METHODS

We set up a clinical service allowing appropriate PH patients to be referred to us for an ICM (LinQ-Medtronic) for diagnosis and monitoring. Abnormal rhythms detected or symptomatic episodes are recorded by the device and a home monitor downloads daily to a secure server accessible by the clinical team.

A PH-specific treatment pathway was developed by the PH team and the cardiology team to standardise treatment for these patients. The pathway advises on anticoagulation, most appropriate antiarrhythmic drug use and allows access to the heart rhythm MDT and consideration for early catheter ablation.



## RESULTS

October 2019 to April 2021 42 patients with PH has an ICM implanted.

### Demographics

Age	50± 14.5
Female	33 (79%)
Caucasian	37 (87%)
Idiopathic PAH	42 (100%)
NO responder	9 (21%)
Mutation positive	10 (24%)
WHO FC I	1 (2%)
WHO FC II	17 (40%)
WHO FC III	21 (50%)
WHO FC IV	3 (7%)
NT Pro BNP	1353±3001
Dual oral therapy	16 (38%)
Triple oral therapy	5 (12%)
Oral + nebs	5 (12%)
Oral + iv prostanoid	11 (26%)

NSR on baseline ECG	40 (95%)
Previous symptoms of arrhythmia	15 (36%)
Previous diagnosis of arrhythmia	3 (7%)
On antiarrhythmic drugs at baseline	7 (17%)

So far we have collected 40.7 patient years of heart rate, heart rate variability, arrhythmia and physical activity data.

### New arrhythmias diagnosed by ICM monitoring

New diagnosis	Number of patients	Outcome
Atrial Flutter	3	Medication + Flutter ablation
Atrial tachycardia	2	Monitoring
Symptomatic frequent atrial ectopy	2	Observe
Atrial Fibrillation	1	Medication + DCCV
Symptomatic inappropriate sinus tachycardia	1	Observe
2:1 Heart block with syncope	1	Dual chamber pacemaker

## Patient example 1

61 year old female, IPAH with previous syncopal episodes and Holter monitoring demonstrating normal sinus rhythm (NSR) therefore deemed to be PAH-related and vasodilator therapy was escalated accordingly. An ICM was implanted in February 2020. The patient re-presented in 2021 with another syncopal episode. Her 12-lead ECG demonstrated normal sinus rhythm but review of her ICM data demonstrated that the syncopal episode coincided with an episode of 2:1 heart block (previously undiagnosed). A dual chamber pacemaker was implanted locally.

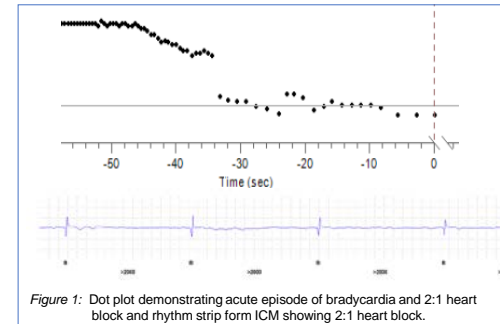


Figure 1: Dot plot demonstrating acute episode of bradycardia and 2:1 heart block and rhythm strip from ICM showing 2:1 heart block.

## Patient example 2

ICM implanted in a 24 year old patient with IPAH and severe RVSD. She was diagnosed with haemodynamically stable atrial flutter and commenced on medical therapy and a DOAC with an aim to perform cardioversion a month later. Her ICM demonstrated she had reverted to NSR and her cardioversion was cancelled. A few weeks later she became increasingly SOB with reduced ET and increase in pedal oedema. An ICM download demonstrated recurrent atrial flutter. She was admitted for diuresis and rhythm control and was referred for a flutter ablation which was acutely successful.

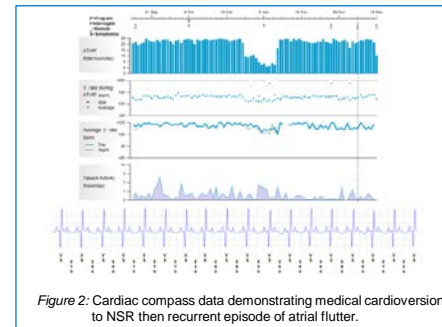


Figure 2: Cardiac compass data demonstrating medical cardioversion to NSR then recurrent episode of atrial flutter.

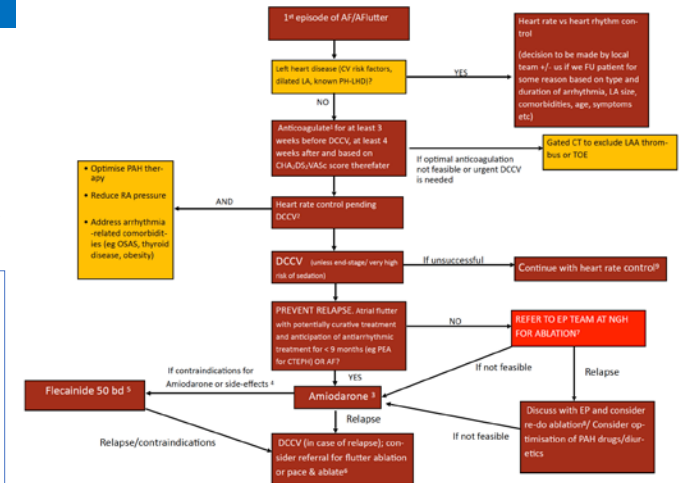


Figure 3: PH-specific treatment pathway designed for Sheffield Teaching Hospitals. J.Middleton, A.Charalampopoulos, A.Kyriacou, A.Rothman

## DISCUSSION

The use of ICM technology in our PH population has allowed continuous, remote monitoring of symptomatic patients referred to Sheffield Pulmonary Vascular Disease Unit. Symptoms are correlated to ICM data enabling quick diagnosis of arrhythmia or symptoms secondary to PH. Patients diagnosed with an arrhythmia now have a PH-specific treatment pathway enabling streamlined, guideline-directed treatment.

## CONCLUSIONS

We have successfully set up a clinical ICM service for PH patients in Sheffield enabling an increased diagnostic certainty for symptomatic patients. Those diagnosed with an arrhythmia are no treated using a PH-specific treatment algorithm enabling safe, guideline-directed management and access to specialist cardiology care.

## REFERENCES

- 1 Galie et al. Guidelines for the diagnosis and treatment of pulmonary hypertension. *ERJ*.2009;34(6):1219-63.
- 2 Brugada.J et al. ESC guidelines 2019/ Supraventricular Tachycardia 9for the management of patients with). *EHJ*: 2019.