

**Supplementary data:**

**Supplementary table 1: Cardiac Magnetic Imaging at presentation in 2014**

<b>Left Ventricular End Diastolic Volume (mL)</b>	131
<b>Left Ventricular End Systolic Volume (mL)</b>	63
<b>Left Ventricular Stroke Volume (mL)</b>	68
<b>Left Ventricular Ejection Fraction (%)</b>	52
<b>Right Ventricular End Diastolic Volume (mL)</b>	137
<b>Right Ventricular End Systolic Volume (mL)</b>	72
<b>Right Ventricular Stroke Volume (mL)</b>	65
<b>Right Ventricular Ejection Fraction (%)</b>	47

**Supplementary table 2: Right Heart Catheterizations at serial time points**

	<b>At presentation (2014)</b>	<b>Under PAH specific medication (2016)</b>	<b>After stopping PAH specific medication (2016)</b>
<b>Systolic Pulmonary Artery Pressure (mmHg)</b>	82	56	65
<b>Diastolic Pulmonary Artery Pressure (mmHg)</b>	40	23	30
<b>Mean Pulmonary Artery Pressure (mmHg)</b>	56	37	42
<b>Mean Right Atrial Pressure (mmHg)</b>	13	11	6
<b>Arterial Blood Pressure (mmHg)</b>	92/64	100/56	106/77
<b>Cardiac Output (l/min)</b>	6.5	13.0	8.3
<b>Pulmonary Vascular Resistance (dyn·s/cm<sup>5</sup>)</b>	492	176	270
<b>Systemic Vascular Resistance (dyn·s/cm<sup>5</sup>)</b>	742	367	777
<b>Pulmonary Arterial Wedge Pressure (mmHg)</b>	16	15	15
<b>Mixed Venous Oxygen Saturation (SvO2 %)</b>	82	87	87
<b>Arterial Oxygen Saturation (SpO2 %)</b>	96	98	99
<b>New York Heart Association (NYHA) Class</b>	3	1	1

**Supplementary table 3: Laboratory results at presentation and during treatment**

	<b>At presentation (2014)</b>	<b>During PAH specific treatment (2016)</b>
<b>Haemoglobin (g/dL)</b>	16.0	17.1
<b>Haematocrit (%)</b>	57	57
<b>Mean corpuscular volume (fl)</b>	72	
<b>Thrombocytes (x 10<sup>9</sup>/L)</b>	118	101
<b>Leukocytes (x 10<sup>9</sup>/L)</b>	4.6	3.8
<b>Reticulocytes index (%)</b>	18	
<b>Erythropoeitin (mIU/ml)</b>	9232	18600
<b>Bilirubin (umol/L)</b>	22	9
<b>Alkaline phosphatase (U/L)</b>	71	54
<b>Gamma glutamyl transpeptidase (U/L)</b>	30	26
<b>Lactate dehydrogenase (U/L)</b>	546	302
<b>Iron (umol/L)</b>	6.2	6.0
<b>Transferrin saturation (g/L)</b>	2.3	2.2
<b>Ferritin (ug/L)</b>	7	11
<b>Creatinin (mmol/Lumol/L)</b>	44	47
<b>NT-pro-BNP* (pg/mL)</b>	196	155

**\*N-Terminal of Brain-type Natiuretic Peptide**