

pathogenesis of PH associated with SCD. The roles of IL 8 and SCF warrant further investigation.

**P36 FUNCTIONAL CLASS AND GAS TRANSFER ARE USEFUL TOOLS IN THE ASSESSMENT OF PULMONARY HYPERTENSION ASSOCIATED WITH SICKLE CELL DISEASE**

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**Introduction and Objectives** WHO Functional class (FC) is an important tool in the management of patients with pulmonary arterial hypertension. There is an increasing awareness of pulmonary hypertension (PH) as a complication of Sickle cell disease (SCD). We wished to evaluate functional class and gas transfer (diffusion capacity DLCOc, KCOc) in patients with SCD (HbSS) with and without PH based on trans thoracic echocardiogram.

**Methods** 32 patients were reviewed and their functional class was determined. Trans thoracic echocardiograms were reviewed in these patients for evidence of PH (defined as a tricuspid regurgitation jet velocity TRV  $\geq 2.5$  m/s). Haematological and lung function data were also reviewed. Only patients in steady state and without a crisis in the preceding 2 weeks were included.

**Results** Of patients with PH 36% (5/14) were FC  $\geq 3$  compared to 6% (1/18) without PH. There was a significant difference in functional class in patients with and without PH on echocardiogram ( $p < 0.05$ ). There was a significant correlation between TRV and WHO functional class ( $p = 0.002$ ,  $r = 0.54$ ). The correlation between TRV and functional class remained significant after correction for haemoglobin. Mean haemoglobin was  $8.3 \pm 1.3$  g/dl in those with PH and  $10.1 \pm 1.4$  g/dl in those without PH ( $p = 0.001$ ). Neither PH nor FC was associated with parenchymal abnormalities visible on CT and/or chest radiograph. PH was also associated with a lower diffusion capacity (DLCOc mmol/min/kPa:  $3.60 \pm 0.79$  vs  $7.60 \pm 2.66$ ,  $p = 0.034$ ) and a lower diffusion capacity corrected for alveolar volume (KCOc mmol/min/kPa/l:  $1.04 \pm 0.31$  vs  $1.96 \pm 0.3$ ,  $p = 0.034$ ). TRV was significantly correlated with DLCOc only ( $p = 0.036$   $r = -0.79$ ).

**Conclusion** Although patients with Sickle cell disease may have a number of reasons to account for their breathlessness, functional class and measurements of gas transfer may be useful markers in screening patients for pulmonary hypertension regardless of low steady state anaemia.

**P37 FLYING—SAFER THAN WE THOUGHT? A QUESTIONNAIRE-BASED STUDY OF 156 INDIVIDUALS WITH HEREDITARY HAEMORRHAGIC TELANGIECTASIA; 95 WITH PULMONARY AVMS**

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**Introduction** Flight has become an integral part of modern life: 65.9 million passengers used Heathrow airport in 2009.<sup>1</sup> Flight, however, exposes us to a unique environment with a reduced  $\text{FiO}_2$ , increased humidity and relative immobility. Although guidelines have been published,<sup>2</sup> there are relatively limited published data regarding flight safety. Patients with particular conditions may provide high risk models for the general population regarding the safety of the flight environment. Here we report the results of a questionnaire-based study in individuals with hereditary haemorrhagic telangiectasia (HHT), a condition that leads to recurrent nosebleeds, but

more importantly for respiratory physicians, frequent pulmonary arteriovenous malformations (PAVMs) that result in a right to left shunt, hypoxaemia, and risk of paradoxical embolic stroke.<sup>3</sup>

**Methods** PubMed searches identified no studies into the effect of flight in HHT or PAVM patients. With ethical approval (NRES 10/H0806/8), patients with definite HHT who had attended our hospital's HHT clinic, were sent a questionnaire. Participants were asked to document flights they had taken, and to list any symptoms they experienced during or shortly afterwards. Responses were correlated with physiological variables in their hospital records.

**Results** 156 replies were received from 308 questionnaires (response rate 50.6%). 145 individuals (95 [65%] with pulmonary AVMS), had flown, reporting 18 943 flight hours in 3950 flights. The median number of flights per patient was 17 short haul, and 8 long haul. There was no difference in erect  $\text{SaO}_2$  between those who reported breathlessness and those who did not (median 93 (range 85–96)% vs median 94 (84–99)% respectively. Individuals who reported breathlessness stated that in-flight oxygen was of benefit (Abstract P37 Table 1).

**Abstract P37 Table 1 All six patients reporting dyspnoea had PAVMs**

	Asymptomatic	Nosebleed	Dyspnoea	Deep venous thrombosis	Ischaemic Stroke
All flights					
Patients (n (%))	111 (77)	26 (18)	6 (4)	2 (1)	1 (<1)
Short haul (<6 h)					
Patients (n (%))	130 (89)	14 (10)	3 (2.1)	0 (0)	0 (0)
Flights taken (n (%))	2864 (96)	78 (3)	39 (1)	0 (0)	0 (0)
Long haul ( $\geq 6$ h)					
Patients (n (%))	120 (83)	20 (14)	6 (4)	2 (1.4)	1 (0.7)
Flights taken (n (%))	837 (86)	112 (12)	17 (2)	2 (<1)	1 (<1)

**Conclusion** These data suggest that even in a population group expected to be more susceptible to complications, flying appeared safe for the vast majority of individuals. A minority of HHT patients did experience adverse symptoms. These were difficult to predict based on clinic measurements.

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**P38 WHICH PATIENTS WITH PULMONARY ARTERIOVENOUS MALFORMATIONS ARE DYSPNOEIC? RETROSPECTIVE ANALYSIS OF A SINGLE CENTRE 2005–2010 COHORT**

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**Introduction/objectives** Pulmonary arteriovenous malformations (PAVMs) are aberrant connections between pulmonary arteries and veins, creating a right-to-left shunt. Hypoxaemia is common,<sup>1</sup> but dyspnoea is usually not the presenting complaint.<sup>2</sup> We hypothesised this may be relevant to dyspnoea mechanisms in the general population.

**Methods** With ethical approval, new patients presenting with CT-proven PAVMs between June 2005 and July 2010 were studied retrospectively. Based on self-reported exercise tolerance at presentation, and blinded to physiological measurements, two