Results 551 responses were received. Participant mean age was 58.3 ± 16.6 years and age at diagnosis was 52.3 ± 18.8 years. 49% of patients had symptoms for >6 months before going to see a doctor and 22% were seen by 4 or more doctors before diagnosis. Time from first symptoms to diagnosis was >1 year in 49% with 31% of patients admitted as an emergency because of their symptoms. After diagnosis, 48% see a specialist at least every 6 months, 87% at least every 12 months. 62% think the support they receive is excellent and 26% good. 90% thought it was better to travel to a Specialist PH Centre rather than to be under the care of a non-PH specialist at a more local hospital.

Discussion This survey found that care of patients with PH is generally good or excellent and patients were keen to travel to Specialist PH Centres for their care. The early symptoms of pulmonary hypertension can be mild and are common to many diseases so it is often a lengthy process to arrive at the diagnosis. Compared with the previous survey the percentage of patients seeing >4 doctors before diagnosis was reduced (22% *vs* 47%) suggesting an increasing awareness of pulmonary hypertension amongst physicians. However, 49% of patients had symptoms for >6 months before presenting, which has not improved since previous surveys. As earlier diagnosis of patients results in better long-term survival, further work should be undertaken to continue to raise awareness in the UK of pulmonary hypertension.

P32 A NEW NURSE LED P.E CLINIC 2015

S Goodman. *University Hospital Southampton, Southampton, UK*

10.1136/thoraxjnl-2016-209333.175

In 2015 Dr R Limbrey (Respiratory Physician) and Sr S.Goodman (CNS Pulmonary Vascular Service) set up a Nurse Led (new patient) Pulmonary Embolism (P.E) clinic to support increasing demand on the traditional model of consultant care.

We aim for a CNS review of all new patients with P.E at 3 weeks following diagnosis, and provide follow up for 2 years. Previously a CNS led follow up clinic was in place.

We aim to provide our patients with early high quality education and information to reduce anxiety, improve physical functioning and quality of life. We suggest that this will reduce perceived ongoing symptoms and lessen follow up requirement.

Referral is made electronically. The referral system has been developed as a learning opportunity to enable medical referrers to identify provoking factors and quantify risk associated with the event. The data obtained from referrals provides support for service provision.

In 2015, 260 patients were reviewed in the Nurse led clinic, 95 of these were new referrals.

Of the 95 new referrals to the service 54 (57%) were provoked by an identifiable transient provoking factor. ESC (2014). Of these 42 (78%) also had persistent risk factors by ESC

Unprovoked group:

The remaining 41 (43%) had no identifiable transient provoking factor. Of these 32 (78%) did have persistent risk factors as per ESC 2014.

We note that 44% of all identified PE patients in this cohort, provoked and unprovoked, had a BMI $\geq$ 30kg/m^2

In the provoked group 16 new patients were reviewed and discharged after an average of 2.62 appointments per person with an additional telephone review planned at the 2 year point.

In the unprovoked group 12 patients have been discharged to date with an average of 3.3 appointments per person with an additional telephone review planned for the 2 year mark.

The remainder of these patients can reasonably be expected to be discharged from follow up with similar levels of review.

Feedback from our patients has been overwhelmingly positive, most commonly expressed as a significant reduction in anxiety and improvement in quality of life.

Abstract P32 Table 1

No of patients seen in Nurse led P.E clinic 2015 by identified transient provoking factors (new patients)	No of patients 54 in total	Persistent provoking factors identified	No of patients
Post operative	17/54	Obesity	26/54
Travel	11/54	Hypertension	12/54
Immobility	8/54	Previous VTE	6/54
Pneumonia	7/54	A.F	2/54
Oral Contraceptive Pill	4/54	Age > 80yrs	2/54
Pregnancy	3/54	Known thrombophilia	2/54
Cancer (identified prior to diagnosis of VTE)	1/	Tamoxifen	0/54
Other	3/54	Diabetes	1/54

Unprovoked group:

Persistent risk factors identified in Nurse led P.E clinic 2015 in Unprovoked group	No of patients
Obesity	16/41
Hypertension	13/41
Previous VTE	7/41
Age > 80 yrs at time of event	8/41
Diabetes	5/41
Known Thrombophilia	2/41
Atrial Fibrillation	1/41
Tamoxifen	1/41

P33 PATIENTS WITH PULMONARY ARTERIOVENOUS MALFORMATIONS AND HEREDITARY HAEMORRHAGIC TELANGIECTASIA REPORT FORCED EXPIRATORY MANOEUVRES DURING PULMONARY FUNCTION TESTS PROVOKE NOSEBLEEDS AND MIGRAINES

[1]HC Tighe, [1]H McKernan, [1]JT Springett, [1]L Babawale, [1]J Perks, [2]T Patel, [3]CL Shovlin. *[1]Imperial College Healthcare NHS Trust, London, UK; [2]St George's Hospital, London, UK; [3]Imperial College London, London, UK*

10.1136/thoraxjnl-2016-209333.176

Introduction and objectives Forced expiratory manoeuvres during lung function testing produce major pressure swings that are often overlooked by referring clinicians. Standard tests use a noseclip to prevent air leakage through the nose. Our goal was to examine how often the tests caused clinical sequelae such as nosebleeds in people with abnormal nasal and pulmonary vasculature due to hereditary haemorrhagic telangiectasia (HHT).

Methods With ethical approval, self-reported migraine features and exacerbations were examined in HHT subjects with and

without pulmonary AVMs, for a series of noninvasive and invasive investigations, using an unbiased online survey.

Results Of 677 HHT-affected participants, 284 reported performing forced expiratory manoeuvres either with or without a noseclip in place. The median age was 54 years (interqurtile range 44–62) and the majority were female. All described nosebleeds: 130 (46%) experienced these at least daily, and a further 95 (33%) at least once per week. Only 1 of 253 (0.4%) reported nosebleeds were worse after finger oximetry measurements, compared to 53/282 (18.8%) after "blowing out hard without a noseclip" (chi-squared $p < 0.0001$). A higher proportion still reported nosebleeds were worse when a noseclip was used (66/192 (34.3%, $p = 0.0003$)). Similarly, migraine headaches (which are more frequent in people with HHT), were reported to be worse after forced expiratory manoeuvres both with (10/85 (11.7%) and without (11/105 (10.5%)) a noseclip, but not after oximetry or being weighed.

Conclusion Noseclip use should be restricted in people already experiencing regular nosebleeds, and further pretest information may be required.

P34 LONG TERM OUTCOMES FOR PATIENTS WITH PULMONARY ARTERIOVENOUS MALFORMATIONS CONSIDERED FOR LUNG TRANSPLANTATION

[1]CL Shovlin, [1]JMB Hughes, [2]M Layton, [1]E Boother, [1]DJ Allison, [2]JE Jackson. *[1]Imperial College London, London, UK; [2]Imperial College Healthcare NHS Trust, London, UK*

10.1136/thoraxjnl-2016-209333.177

Introduction and objectives Pulmonary arteriovenous malformations (PAVMs) are abnormal vessels that provide anatomic right-to-left shunts, and frequently result in severe hypoxaemia. Lung transplantation is sometimes considered if PAVMs are not amenable to treatment by embolization or surgical resection.

Methods A retrospective review was performed of patients with PAVMs assessed between 1999 and 2016 at a single UK institution. Characteristics of those considered and referred for lung transplantation assessment were examined.

Results Between May 1999 and July 2016, 707 patients with PAVMs were reviewed. Six were or had been formally considered for lung transplantation purely for PAVMs, 4 as adults (3 at our institution between 1989–1995), 2 as children. Ages ranged from 2–47 (median 22) years. Four were from the UK, two had been assessed in non UK countries. All had hereditary haemorrhagic telangiectasia (HHT). Three had suffered a cerebral abscess due to their PAVMs. The adults had undergone maximal embolisation of PAVMs, and the children were considered to have untreatable disease. The median SaO^2 was 79% on air.

One adult underwent lung transplantation overseas and died peri-operatively. The other five were not transplanted, in four cases at the patients' request. Currently, their survival post assessment for lung transplant ranges from 15 to 27 (median 22) years, with four of the five still alive.

All five maintained arterial oxygen content by marked secondary erythrocytotic responses when not iron deficient. Four had subsequent embolisation treatments. One had three successful pregnancies, and another is working full time with no supplementary oxygen. However, two had a cerebral abscess (one fatal); three had deep vein thromboses; two experienced long term disabling pain attributed to hypertrophic pulmonary osteoarthropathy in one; and three have exercise limitation requiring supplementary oxygen. Antiplatelet therapy was recommended for two due to ischaemic strokes; one remains anticoagulated 16 years after the insertion of a caval filter; and one is regularly evaluated for hyperviscosity symptoms that would precipitate isovolaemic venesection.

Conclusion Very long term stability is evident in the five patients who did not receive a lung transplant. The cohort remain at high risk of cerebral abscess and other complications of right-to-left shunting.

P35 A RETROSPECTIVE OBSERVATIONAL STUDY DESIGNED TO CHARACTERISE INDIVIDUALS WITH PULMONARY ARTERIOVENOUS MALFORMATIONS (PAVMS) AND CEREBRAL ABSCESSES AT A SINGLE INSTITUTION

[1]EJ Boother, [2]S Brownlow, [2]JE Jackson, [3]CL Shovlin. *[1]Imperial College School of Medicine, London, UK; [2]Imperial College Hospital NHS Trust, London, UK; [3]Imperial College London and Imperial College Hospital NHS Trust, London, UK*

10.1136/thoraxjnl-2016-209333.178

Introduction and objectives Individuals with pulmonary arteriovenous malformations (PAVMs), who often concurrently have hereditary haemorrhagic telangiectasia (HHT), are at increased risk of cerebral abscess, frequently resulting in life-changing deficits. Current aetiological consensus supports paradoxical emboli of periodontal origin, with preventative dental recommendations published in 2008.[1] Limited knowledge is available to identify specifically at-risk individuals. This study aimed to characterise a cohort from 2005–2016; recognise potential precipitants; and compare results to an earlier published series.

Methods With ethical approval, notes of patients newly presenting at a single institution between 2005–2016 were reviewed to collect cohort characteristics. GraphPad Prism was used to calculate descriptive statistics, and to perform Mann-Whitney and Chi-squared statistical tests for comparison between the two cohorts.

Results Of 488 new patients with PAVMs, 33 (6.8%) had cerebral abscesses. 21 were female (63.6%), 12 male. The rate corrected for ascertainment bias was 3.8%. The median age at abscess was 46 years (range 13–69). The median oxygen saturation (SaO_2) was 90.75% (range 70–97.5%), with 9 individuals having respiratory symptoms. There were 29 confirmed HHT diagnoses (87.9%). The median largest feeding artery diameter was 5mm, and for 5 individuals, all feeding arteries had diameter ≤3 mm. In total, 19 (57.6%) had residual PAVMs too small for embolization.

Organisms identified (Table 1) suggest periodontal origin; 16 individuals (48.5%) had poor dental hygiene and 9 (27.3%) had dental events as abscess precipitants. Interestingly, 4 individuals had abscesses whilst on holiday abroad. 5 individuals reported worsening migraines and 2 individuals had increased seizure frequency ≤ 3 months pre-abscess.

Within the non-overlapping 1999–2005 cohort, there were 28/219 abscesses (12.8%, 9.05% adjusting for ascertainment bias). Compared to the later series, similar proportions of abscesses occurred prior to PAVM diagnosis (18/28 (64.3%) vs. 24/33 (72.3%)). Proportionally more males were affected (57.1%). No other significant differences were found for age, SaO_2, feeding artery diameter, presence of respiratory symptoms, or HHT (all p values >0.12).

Conclusion The PAVM cohort remains at high risk of cerebral abscess. Scrupulous dental hygiene appears to remain paramount to reduce risk. The worsening migraines and abscess occurrence whilst abroad are unexpected findings recommended for further investigation.