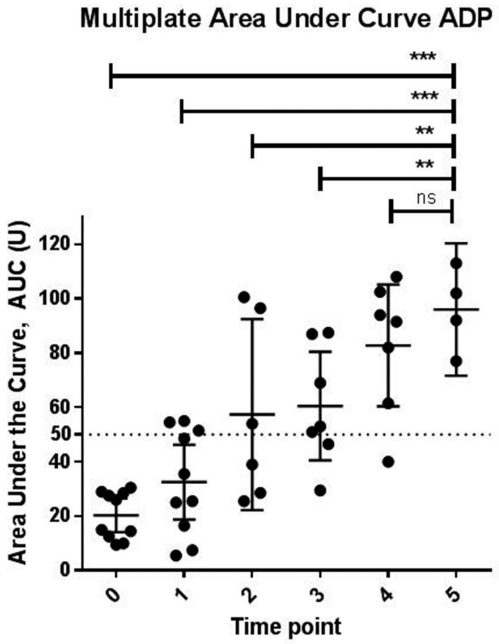
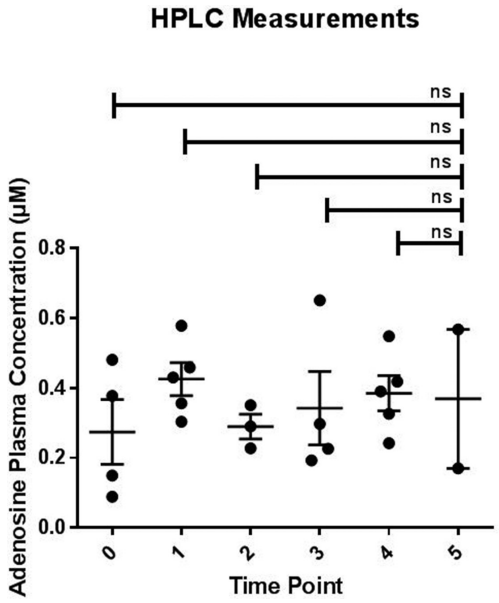


Abstract 72 Figure 1



Abstract 72 Figure 2

Congenital Heart Disease

73 **IMPORTANCE OF CLASSIFICATION AND REASSESSMENT IN PATIENTS WITH PAH-CHD: THE NORPAP DATABASE**

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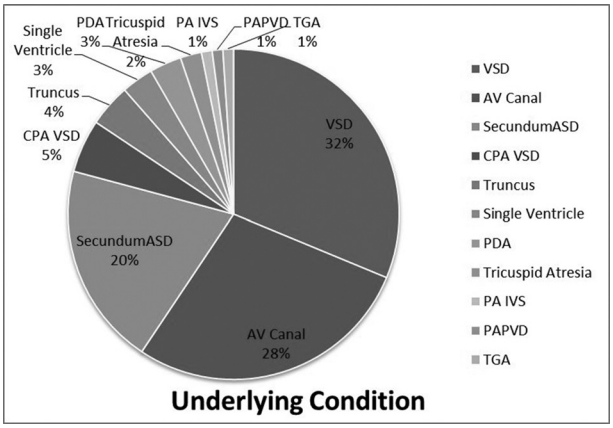
Background Patients with Pulmonary Arterial Hypertension associated with Congenital Heart Disease (PAH-CHD) are subdivided into 4 categories: 1) Eisenmenger Syndrome, 2) PAH with systemic-to-pulmonary shunts, 3) PAH with coincidental defects, 4) PAH after defect correction. There is clear benefit for targeted therapy in Category 1, 2 (less so in those with segmental PAH), and 4.



Abstract 73 Figure 1

Methodology The NORPAP database was created in 1993 and contains 2854 patients. Patients in Norwich and Papworth have access to the joint GUCH-PVDU (Grown Up Congenital Heart Disease – Pulmonary Vascular Disease Unit) service. Patients with PAH-CHD were extracted from the NORPAP database and the above classification system applied.

Results 98/2854 (3.4%) of patients had PAH-CHD. Female: male ratio 1.65:1. Age range 18–96, mean age 47, median 43.5. Trisomy 21 in 40 patients. Underlying condition shown in Figure 1. Number of patients per Category is as follows (Figure 2): Category 1 (71/98=72%, Age Range=25–92, median 42), Category 2 (18/98=18%, Range 18–96, median 58), Category 3 (3/98=3%, Range 38–90, Median 42), Category 4 (6/98=6%, Range 24–69 Median 45: diagnoses – DORV, VSD, TGA (atrial switch), PDA, PA-VSD, secundum ASD). 2 patients in category 4 had mixed aetiology with Chronic Thromboembolic Pulmonary Hypertension (CTEPH).

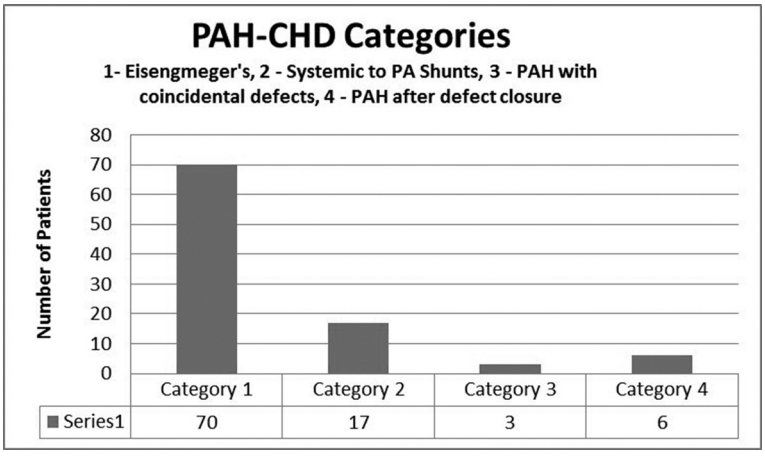
26/98 (27%) of patients have died. Median age at death 41. Category 1: 21/71 29.5%, Category 2: 2/18=11%, Category 3: 1/3=33%, Category 4: 2/6=33%.

55/98 (56%) are on targeted therapy. In the Eisenmenger group 41/71 (58%) are on targeted therapy. Sildenafil 11/41=26.8%, Bosentan 13/41=31.7%, Ambrisentan 1/41=2.4%, two agent therapy 16/41=39%.

Pre-treatment 6 min walk mean 288.5m, post-treatment mean 320.6m. Paired two sample t-test analysis reveals a significant difference in 6 min walk test (t-stat 2.02, p 0.01).

Of note was 28 year old Patient X: Trisomy 21 with large VSD and subpulmonary stenosis and suggestion of double outlet RV on poor quality echocardiography. Reassessed with cardiac MRI which confirmed tetralogy of Fallot. Following RHC, which showed RVOT gradient of 18 mmHg, mean PA pressure 55, PVR 25 wood units falling to 15.8 with maximal pulmonary vasodilatation, he was started on sildenafil.

Conclusion Targeted therapy in PAH-CHD improves quality of life, delays time to deterioration and may have a mortality benefit.Our limited data supports the evidence that targeted therapy improves objective exercise tolerance. Adult follow up surveillance clinics must be alert to deterioration of functional class in Category 1; Trisomy 21 patients that have been labelled as PAH, but may have normal PA pressure at right



Abstract 73 Figure 2

heart catheterisation; high index of suspicion with new symptoms in repaired CHD who develop PAH (Category 4). Although Patient Xs management did not change, we highlight the case to remind clinicians of need to always reassess patients with historical and sometimes inaccurate labels.

74 MULTISYSTEM MANIFESTATIONS OF CYANOTIC ADULT CONGENITAL HEART DISEASE: REVIEW OF PREVALENCE AND CURRENT PRACTICE ON THE NORPAP DATABASE

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10.1136/heartjnl-2017-311726.73

Introduction Adults with congenital heart disease may be cyanotic either from unrepaired cyanotic conditions, or due to the failure of palliative procedures to completely separate the systemic and pulmonary circulations. Clinicians should be aware of the multisystem manifestations of chronic cyanosis.

Chronic hypoxaemia leads to a physiological rise in haemoglobin and red cell mass, causing secondary erythrocytosis. Management includes avoidance of routine venesection, ensuring adequate hydration and avoiding iron deficiency. Complete iron studies should be carried out regularly since the MCV is a poor determinant of iron deficiency.

Methodology The NORPAP database was created in 1993 and contains 2854 patients from Norwich and Papworth. Patients with cyanotic heart conditions were extracted from the database and the prevalence of the following complications were sought from electronic letters: hyperviscosity symptoms, acne, gout, bleeding/haemoptysis, thromboembolism, brain abscess, stroke, endocarditis, renal dysfunction and gallstones (Table 1).

In addition the management of erythrocytosis was assessed as satisfactory vs not satisfactory based on the following criteria annual FBC and complete iron studies including iron supplementation if indicated, and avoidance of routine venesection.

Results 102/2854 (3.5%) had cyanosis. PAH-CHD present in 70/102 (68.6%). F:M 1.76:1. Age range 21–81, mean 39, median 38. Death occurred in 26/102 (25.4%). Mean age at death 45, median 42. Cause of death Cardiac/PAH: 12/26, non-cardiac: 5/26, unknown: 9/26. In the Trisomy 21 group mean age at death 40, median 38.

Underlying condition is shown in Figure 1. One patient with VSD Eisenmenger Syndrome had concomitant ischaemic heart disease, complicated by retroperitoneal bleeding secondary to dual antiplatelet therapy and low molecular weight heparin.

Complete clinical information and blood results were available for 78/102 patients.

Erythrocytosis: 66/78 (84.6%) satisfactory, 12/78 (15.3%) not satisfactory. This was predominantly on account of no iron studies.

Conclusions An understanding of the multisystem manifestations of cyanotic conditions and avoidance of common pitfalls is paramount in the management of these complex patients. We follow the ESC/AHA guidelines on antibiotic prophylaxis against endocarditis in high risk groups. None of the documented cases were related to dental procedures.

Patients need careful assessment when considering anticoagulation given the balance of risk between bleeding and thromboembolism complications seen not infrequently in our population.

Gout and renal dysfunction are common and should be actively sought during routine clinical assessment. On review of the data from this observational study, a clinic proforma to include a checklist of symptoms to screen for, in addition to FBC, Iron Studies, Uric Acid, and U and Es has been developed.

Abstract 74 Table 1

Complications	Number	Percentage
Hyperviscosity Symptoms	4	5%
Acne	6	7.6%
Gout	16	20.5%
Bleeding/Haemoptysis	9	11.5%
Thromboembolism	6	7.6%
Brain Abscess	4	5%
Stroke	3	3.8%
Endocarditis	5	6.4%
Renal Dysfunction	24	30.7%
Gallstones	1	1%