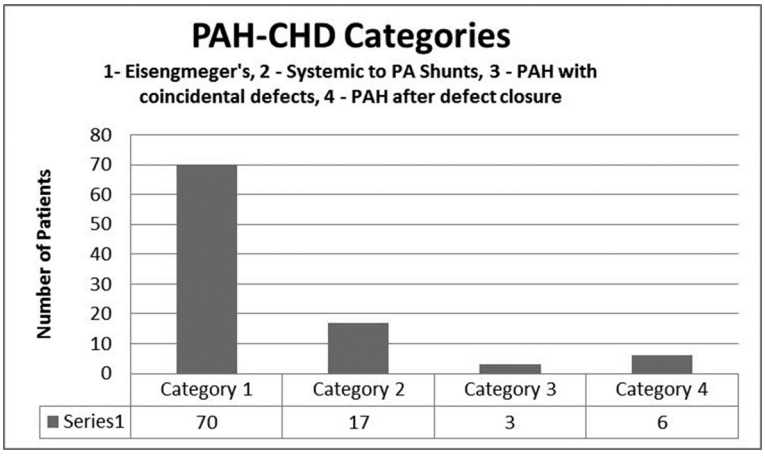

Abstract 73 Figure 2

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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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.

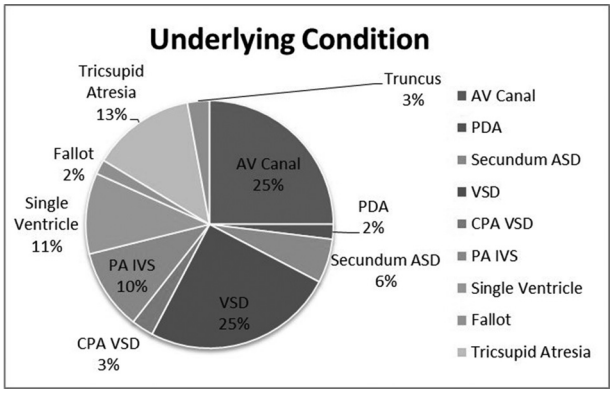
Conclusions An understanding of the multisystem manifestations of cyanotic conditions and avoidance of common pitfalls is paramount in the management of these complex patients. We follow the ESC/AHA guidelines on antibiotic prophylaxis against endocarditis in high risk groups. None of the documented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



Abstract 74 Figure 1

75 INHERITED LEFT VENTRICULAR NON-COMPACTION IN SPONTANEOUS MONOCHORIONIC DIAMNIOTIC TWIN PREGNANCY

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Introduction Pregnancy in Left Ventricular Non-Compaction (LVNC) can increase the risk of heart failure, arrhythmias and systemic embolic events. LVNC carries an autosomal dominant trait and incomplete penetrance. This case study demonstrates the importance of including genetic inheritance within pre-pregnancy counselling, multidisciplinary care and early detection of clinical deterioration in women with LVNC. Whilst we believe this is the first case study with identified genetic testing with an alteration in gene MYH7.

Case report A 27 year old female with a pre-existing diagnosis of LVNC was referred to the high risk pregnancy service at the University Hospital Southampton, from out of region, at 23 weeks gestation with spontaneous monochorionic diamniotic twins. Initial transthoracic echo (TTE) at 23 weeks gestation demonstrated a dilated and heavily trabeculated left ventricle (LV) at 5.6 cm with severe LV systolic dysfunction, ejection fraction (EF) 20%–25%. The referral pre-pregnancy EF was quoted at 30%–35%. Medical therapy was up-titrated (beta-blockade and diuretics) and patient remained NHYA II.

At 27 weeks gestation the patient presented with a nocturnal cough. Clinical examination demonstrated an elevated jugular venous pressure, worsening mitral and tricuspid regurgitation, peripheral oedema and S_3. Repeat TTE demonstrated a further deterioration in LV function EF 10%–15%, with worsening tricuspid regurgitation and elevated pulmonary artery pressure. The patient was admitted for intravenous diuretics and medication optimisation.

Fetal echocardiogram confirmed the monochorionic twins had inherited the left ventricular non-compaction.

The patient was delivered at 30 weeks by caesarean section in cardiac theatres under general anaesthetic with vascular access in place to allow prompt E-CRP if required.

Postnatally, the patient returned to NYHA II with a 8 month post partum left ventricular EF 30%–35%.

Day 1 TTE of the monochorionic twins confirmed LVNC with identical appearances to their mother (Figure 1). Genetic testing identified an alteration in gene MYH7 (c.3908G>C p.(Arg1303 Pro)) in our patient and her monochorionic twins. This genetic change has not been reported before in other individuals.

Conclusion Novel Genes

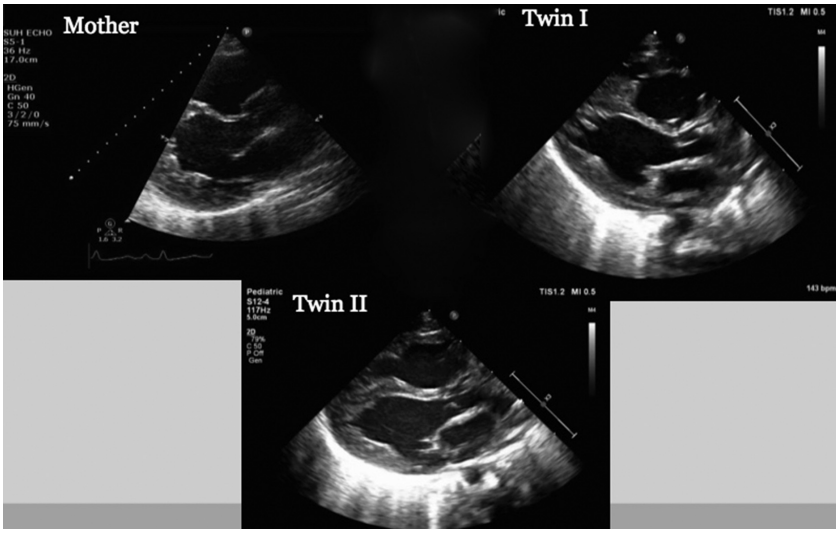
Our patient is the first reported case study of an alteration in gene MYH7 (c.3908G>C p.(Arg1303 Pro)) in LVNC and this alteration has also been identified in her monochorionic twins.

Pre-pregnancy Counselling

This case demonstrates the importance of pre-pregnancy genetic counselling in inheritable dilated cardiomyopathies.

Multidisciplinary Team Work

Our case illustrates the importance of multidisciplinary team working in complex cardio-obstetric cases to ensure the safest outcome for both mother and neonate. Our team included cardiologists, obstetrics, obstetric anaesthetists, cardiac anaesthetists, cardiac surgeons, referring transplant physicians, midwives, cardiac nurses, neonatologists and clinical genetics.



Abstract 75 Figure 1 Plax echocardiographic images of patient (mother) and monochorionic twins