Results We identified 8 patients (6 female) with mean age 26.3±6.2 years. Mean follow-up was 4.1 years (range, 18 months to 12 years). All patients have had surgical repair; one patient required concomitant mitral valve repair. Mean age at operation was 9.7 years (range, 3 months to 34 years old); 3 patients (38%) were operated as adults. At last clinic review, one patient had NYHA III symptoms, the rest were well (NYHA I). Mean peak oxygen consumption on cardiopulmonary exercise testing was 36±2.4 ml/kg/min (range 33.4 to 38.8 ml/kg/min, mean 93%±15.9% predicted, range 78% to 112%). All patients were in sinus rhythm and no ischaemia or arrhythmias were identified. 7 patients (88%) had good left ventricular function (mean EF 61%); 1 patient had mildly impaired function (EF 50%) due to an apical transmural infarction. Moderate mitral regurgitation was seen in 3 patients (38%) and all had normal aortic root size.
Conclusion We describe long-term outcomes of patients with ALCAPA syndrome. Postoperatively, the majority remain asymptomatic with good exercise capacity. Their left ventricular systolic function is good. Life-long follow up is warranted.

10 LONG-TERM OUTCOMES OF ADULTS WITH WILLIAMS SYNDROME IN AN ADULT CONGENITAL HEART DISEASE CENTRE

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10.1136/heartjnl-2017-311499.10

Background Williams syndrome, is a congenital, multisystem disorder, involving the cardiovascular, connective tissue and central nervous systems. We sought to determine long-term outcomes of these patients in our Adult Congenital Heart Disease Service.
Methods A retrospective review of case notes of patients with Williams syndrome was performed, reviewing clinic letters, operation details and cardiac imaging.
Results We identified 37 adults with Williams syndrome (62% male) with mean age 30±9.6 years (range, 19 to 56 years). 6 patients (16%) were discharged after their first review as no cardiovascular abnormalities were identified. One patient was lost to follow-up. Mean follow-up of the rest was 7.9±4.6 years (range, 6 months to 15 years). Most common cardiovascular manifestations included systemic arterial hypertension (40%, n=12), supravalvar aortic stenosis (57%, n=17; repaired (n=10)), pulmonary artery stenosis (30%, n=9; operated (n=5)) and aortic coarctation (20%, n=6; repaired (n=4)). At last clinic review, 10 patients (33%) were NYHA II (n=9) or NYHA III (n=1). 12 patients (40%) were hypertensive (BP >140 systolic), despite being on antihypertensive treatment (n=8). All patients were in sinus rhythm and no arrhythmias were identified. 9 patients (30%) had prolonged QTc (>440 ms in men or >460 ms in women). The majority (97%, n=29) had good left ventricular function (mean EF 63%±4.6%), and only one patient had mild impairment (EF 50%). No significant post-operative gradients were measured.
Conclusion Long-term follow-up of patients with Williams syndrome and significant cardiovascular disease is essential with particular care to blood pressure control.

11 PERSONALISED WARFARIN DOSING IN CHILDREN AFTER CONGENITAL HEART SURGERY: A RANDOMISED, PROSPECTIVE, CROSS-OVER, PILOT STUDY AT GLENFIELD HOSPITAL, LEICESTER

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10.1136/heartjnl-2017-311499.11

After cardiac surgery (eg, Fontan repair, valve replacement), children are at an increased risk of thrombosis and therefore long-term oral anti-coagulation is essential to prevent morbidity and mortality. Warfarin is commonly used, but optimising the dose and maintaining a therapeutic INR is challenging for clinicians due to considerable inter- and intra-individual variability in its pharmacokinetics (PK) and pharmacodynamics (PD). The PK/PD is affected by both patient related (such as genetic polymorphisms of the metabolic enzymes) and environmental (eg, diet) factors. To improve the accuracy and consistency of warfarin dosing, a novel PK/PD (pharmacological) based model incorporating pharmacogenomics has been developed to assist clinicians in predicting initial and maintenance warfarin doses in post-operative cardiac children.[1]

The aim of the study is to compare warfarin dose management using pharmacological model with the traditional, 'trial and error' approach. The study is prospective and observational and involves 2 groups: In Group 1 (warfarin naïve) patients, loading and maintenance warfarin doses are estimated using the pharmacological model over 6 month duration and compared to historical case matched controls dosed according to the traditional approach. Group 2 patients already established on maintenance warfarin therapy entered a randomised cross-over study comparing pharmacological model-estimated dose adjustments with the traditional approach, over a 12 month period. The study also seeks to explore the views of children, parents and medical staff about the new model based approach.

The study commenced in October 2015 and recruitment stopped in December 2016. Group 1 (n=5) and Group 2 (n=29) participants are currently being followed up for their warfarin dosing and monitoring.

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12 IS ECHOCARDIOGRAPHY ALONE SUFFICIENT FOR RELIABLE CASCADE SCREENING OF FIRST DEGREE RELATIVES OF PATIENTS WITH BICUSPID AORTIC VALVES?

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10.1136/heartjnl-2017-311499.12