patch. A cystic lesion (4.5×4.5 mm) was present in the same position as the original haematoma, suggestive of false aneurysm into which there was arterial flow.

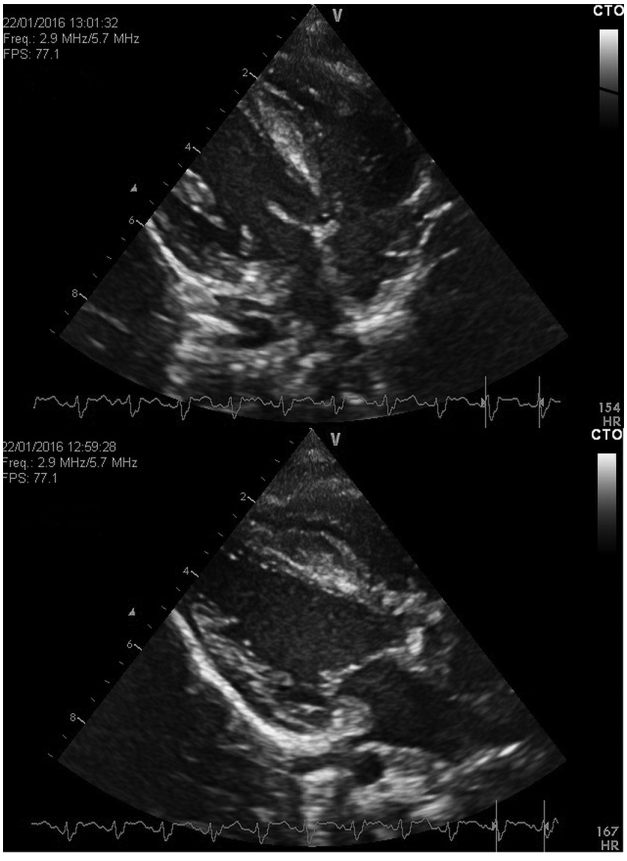
Case two A 10-month-old infant underwent elective repair, following postnatal diagnosis of VSD and mild mitral valve regurgitation. Examination confirmed normal heart sounds with a harsh pansystolic murmur loudest at the left lower sternal border with no respiratory distress and no hepatomegaly.

The defect was a moderate size perimembranous VSD partially covered by the septal leaflet of the tricuspid valve. It was closed with a bovine pericardial patch and 6/0 surgilene running suture. This was supported with a piece of autologous pericardium on the tricuspid valve septal leaflet. An additional two interrupted 6/0 pericardial sutures were placed to secure the patch in position. The infant came off bypass smoothly in sinus rhythm.

No TOE was performed; however, postoperative TTE showed no residual VSD with good ventricular systolic function and an IVS haematoma measuring 11×25 mm. Repeat TTE the following day showed the septal haematoma resolving and less echogenic in appearance. The patient remained haemodynamically stable with no rhythm disturbance and was discharged on Day 6 postoperatively.

On follow-up a month later, he was clinically well; the echocardiogram showed the haematoma had significantly reduced in size (Figure 2A and B). Unlike the previous case, there was cystic collection, and follow-up was arranged for 6 months.

Discussion Haemorrhagic dissection of the IVS after patch closure of a VSD is a rare complication.[1] This is the first report of a case where the haematoma was seen in a patient following ToF repair.



Abstract 8 Figure 2

In both cases, we have described a rapid postoperative increase in the diameter of the IVS which suggests surgical trauma of the coronary septal branch during placement of the septal patch sutures. Anticoagulated, the bleeding probably dissects along a plane beneath the endocardium, resulting in a haematoma that bulges into the ventricular cavity,[2] confirmed by echolucency on postoperative echocardiogram. Follow-up echocardiogram in Case 1 lends support to this theory with coronary flow seen in the cystic lesion within the resolving septal haematoma which likely dissipates into a ventricle.

Most reports are in the adult population where IVS haematoma has been described after myocardial infarction, percutaneous coronary artery intervention and coronary artery bypass grafting.[2] The use of intraoperative TOE is essential in congenital heart surgery to facilitate the early detection of any complications.[2,3] Haemodynamic instability dictates management with prompt reintervention, carried out in those cases with compromise through surgical drainage[4,5] or with needle puncture.[1] The vast majority of adult cases opt for aggressive intervention, thereby potentially influencing decision-making in paediatric cases following surgery.

Myocardial haematoma formation may cause haemodynamic instability or conduction abnormalities that may lead to serious short-term complications, including heart block, outflow obstruction and tamponade.[3] Conservative approach is rarely adopted; however, in haemodynamically stable cases, complete and spontaneous resolution of the haematoma has been seldom demonstrated.[2,4]

Management is dictated by initial presentation, ECGs are mandatory initially to ensure no AV block and as the haematoma resolves, we feel it is unlikely to result in later AV nodal issues. In the absence of haemodynamic instability, echocardiography remains the mainstay of follow-up.

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9 LONG-TERM OUTCOMES OF ADULTS WITH ALCAPA REPAIR

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10.1136/heartjnl-2017-311499.9

Background Anomalous left coronary artery from the pulmonary artery (ALCAPA syndrome) is a rare congenital abnormality, which, if untreated, can cause complications such as myocardial infarction, heart failure and death; only few untreated patients survive to adulthood. 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 ALCAPA was performed, reviewing clinic letters, operation details, cardiac imaging and exercise tests.

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