Objectives The Fontan procedure remains the predominant palliative approach for a single ventricle circulation despite many concerns about long-term elevation of systemic venous pressures on multiple organ systems. Preservation of cardiac function is a prerequisite for health of the Fontan circuit and loss of sinus rhythm with need for implantation of a permanent pacemaker remains a concern. We sought to compare early and late survival, complications and need for further intervention between those patients who require a permanent pacemaker and patients who remain in sinus rhythm following the Fontan procedure.
Methods This single centre, retrospective case note analysis study involved all patients who have undergone the Fontan procedure between 1987 and 2015 and have had regular follow up at Leeds General Infirmary. 145 patients were identified of which two were excluded for having a pacemaker prior to surgery. Of the remainder, 21 patients required a pacemaker. The primary outcome was survival and the secondary outcomes were early and late complications, need for further intervention and oxygen saturation in long-term follow up.
Results There was no difference in survival at 7 days, 30 days, 1 year or 5 years (30 day survival SR 92.6%, non-SR 90.5%, p=0.66). The pacemaker group were more likely to have cerebral or renal complications in the first year post procedure but there was no difference in long term complications such as protein losing enteropathy or pulmonary hypertension (acute kidney injury SR 0.8%, non-SR 19.1%, p=0.002). Patients in the pacemaker group were more likely to undergo further interventions and these interventions were more likely to involve cardiopulmonary bypass (SR 0.06%, non-SR 0.48%, p<0.001). There was no difference in saturations between the two groups in long-term follow up.
Conclusions This study demonstrates the likely cause of loss of sinus rhythm being more complex surgery performed as part of the Fontan procedure. With more active surgical management of these issues in early childhood as well as preference for an extracardiac conduit loss of sinus rhythm is likely to become a less frequent issue. Despite an increase in early complications and the need for further interventions, pacemaker requirement does not affect long term outcome of the Fontan procedure.

16 MANAGING SCIMITAR SYNDROME: THE BIRMINGHAM PERSPECTIVE

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Aim To evaluate the long-term outcomes of patients diagnosed with Scimitar syndrome in Birmingham from 1988–2016.
Methods Data was collected retrospectively using medical records. Scimitar syndrome was defined as a right hemi anomalous pulmonary venous connexion to the Inferior Vena Cava.
Results Twenty-nine patients diagnosed with Scimitar syndrome were identified. Three distinct subgroups emerged: the "Early Scimitars" (n=6), the "Stable Scimitars" (n=11) and the "Surgical Scimitars" (n=12). The Early Scimitars had a median age at diagnosis of 30 days. They all presented with severe respiratory distress and most had associated congenital defects (n=5). There was 100% mortality in this group. Stable Scimitars had a median age at diagnosis of 11 months. In this group, six patients (54.5%) were symptomatic. Coil embolisation of the collateral arterial supply to the right lung abolished symptoms in 50% of cases (n=3). Surgical Scimitars had a median age at diagnosis of 17.3 years; nine (75%) underwent surgical repair of their Scimitar vein. One was lost to follow up. Overall, four patients (50%) developed post-operative complications and two developed Scimitar vein stenosis (25%). There was one late post-operative death (13%).
Conclusion There is a high mortality in the Early Scimitars. Stable Scimitars may benefit from coil embolisation and should be conservatively followed up. Due to the significant risk of complications, surgical correction should be reserved for the symptomatic and those developing co-morbid disease (increasing right ventricular dilation, arrhythmia). If asymptomatic, surveillance in the form of MRI, cardiopulmonary exercise tolerance and echocardiography should be offered.

Abstract 16 Table 1 Summary table of the main characteristics and management approach taken in the different sub-groups of Scimitar syndrome identified.

The Early Scimitars	• Present as neonates • Additional complex anomalies common	**High mortality**
The Stable Scimitars	• Present in early childhood • Significantly hypoplastic R lung • Insignificant L to R shunt	**Benefit from coil embolisation;** conservative follow up
The Surgical Scimitars	• Present in childhood/adulthood • Good R lung • Significant L to R shunt.	**Offer surgery if:** symptomatic, increasing RV dilation, arrhythmia **Surveillance if:** asymptomatic (MRI, CPEX, echo)

17 BURDEN OF ARRHYTHMIAS IN NON-SURGICAL UK ADULT CONGENITAL HEART DISEASE (ACHD) CENTRES: THE NORPAP DATABASE

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Introduction Adults with congenital heart disease have a high burden of arrhythmias related to the underlying substrate, and the consequences of palliation. The NORPAP ACHD database, established in 1993, contains 2587 patients in 2 non-surgical centres.
Methodology Patients with documented arrhythmias were extracted from the NORPAP database and analysed for demographics, underlying condition, surgical intervention, rhythm, and electrophysiological (EP) procedures between 1993 and 2015.
Results 13.6% of patients (353/2587) had documented arrhythmia, mean age 51 years, 50% male, prior surgical intervention in 77.9%. Atrial tachyarrhythmias predominate: Atrial Fibrillation (AF) 35.6%, Atrial Flutter (AFL) 24.3%, Supraventricular tachycardia (SVT) 19.5% and Atrial Tachycardia (AT) 8.2%. It was common for patients to experience more than one episode and type of arrhythmia. High grade AV Block in 8.5%, Sinus Node Dysfunction 3.4%. Ventricular Tachycardia (VT) 9.9%, Ventricular Fibrillation 0.6%. 32% of patients with arrhythmias had EP procedures, 82.4% had prior surgical intervention. Of the total, 88 had RF Ablation; 18%

required repeat procedures. Underlying condition was diverse (21 conditions): Secundum ASD 19/88 (21.6%), Fallot's Tetralogy 19/88 (21.6%), and Ebstein's anomaly 9/88 (10.2%) were the most common.

Conclusions Whilst arrhythmias are most common in Secundum ASD, complex ACHD (ccTGA, complex congenital, TGA and Ebstein's anomaly) have a disproportionately high arrhythmia burden. 10% increase in EP procedures noted compared to our 2006 study. This is appropriate for to avoid recurrent admissions and long term anti-arrhythmics. Complex anatomy, challenging venous access and scar related re-entry tachycardias underline the need to expand the subspecialist EP-CHD capacity internationally.

18 KAWASAKI DISEASE- UNUSUAL FULMINANT PRESENTATION IN A SIX WEEK OLD INFANT WITH SYSTEMIC AND CARDIAC INVOLVEMENT, DIAGNOSIS AND MANAGEMENT CONUNDRUMS

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A six week infant was admitted with three days history of coryza and high grade temperature. She received treatment as presumed sepsis after sending full septic screen, congenital viral and retroviral serologies. She had high inflammatory markers along with raised liver enzymes and thrombocytopenia. She did not return any positive microbiology as well as viral serologies apart from persistently high inflammatory markers.

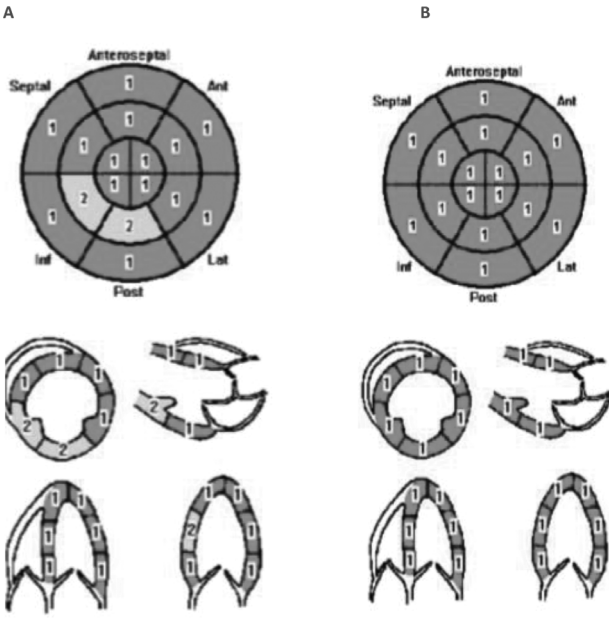
On day five of illness, she developed vasculitic changes in eyes, mouth and extremities concurrently with infiltrative pulmonary disease which required non invasive ventilatory support, hepatitis, renal dysfunction, anaemia, hypoalbuminaemia, hypofibrinogenaemia, raised ferritin, raised triglycerides in addition to thrombocytopenia. Initial echocardiogram was normal.

She was transferred to a tertiary paediatric centre for further support with rheumatology and immunology teams. She had been started on treatment for Kawasaki disease with aspirin and immunoglobulins. A repeat echocardiogram at day seven of illness showed evidence of coronary artery dilatation.

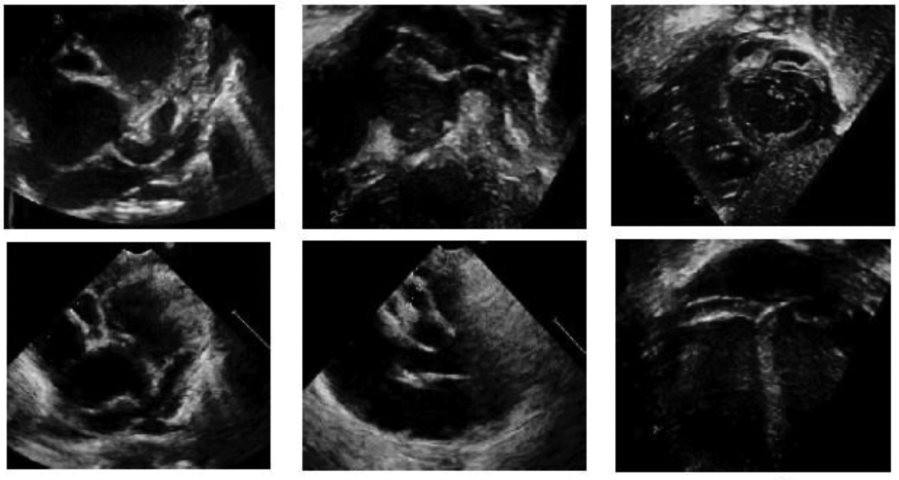
In view of ongoing severe inflammatory changes at two weeks of illness, she received a further course of immunoglobulins along with a course of methylprednisolone and then infliximab afterwards.The coronary artery dilatation had worsened with Z scores of 9–10 in all three coronaries. The skin biopsy was done because of profound vasculitic changes and peeling skin, although showed abnormalities, was not conclusive.

She was then transferred to the intensive care unit in our regional cardiac centre where she was treated with therapeutic heparin infusion then subcutaneous heparin and aspirin and further stabilised before she was successfully discharged home.

Kawasaki treatment guidelines in refractory disease has been updated in our unit.



Abstract 18 Figure 2 A&B Wall motion abnormality scoring 7 weeks (A) and 15 weeks (B) after peek presentation (A).



Abstract 18 Figure 1 Echocardiogram images of Coronary Arteries @ at peak of presentation